NEWSLETTER - AONEI

DARPAN: A Reflection of AONEI Activities

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AONEI Annual Conference (Imphal)
(From left to right)
Dr. L Jaichand Singh, Dr. S Rita Devi (Director, RIMS, Imphal) ,
Dr. H Nandakumar Sharma (Vice Chancellor, Manipur University) , Dr. K Ahmed, Dr. Y Indibor Singh

GUEST COLUMN

BEYOND TREATMENT

On Support Groups - What It Offers Those Faced With a Diagnosis of Cancer

Viji Venkatesh

Region Head (India & South Asia), The Max Foundation | Managing Trustee - Friends of Max | Trustee - Being Human

A diagnosis of cancer brings along with it a great many social, cultural and financial ramifications ; not just to the patient but to the whole family and extended circle of friends and others. It is a life altering event and calls for management on many levels .

Let us look at life as such : Life is a journey and we are all in it together from birth till end of life. For better or for worse.

Of course by large we are not too concerned about end of life are we ? We know it is certain but we do have this precious gift of life with us to cherish and nurture in the meantime . So along with our family, friends , neighbours and colleagues we happily continue on this journey which is full of love , happiness, some challenges, a few or many as the case may be , achievements, progress and success.

But for some of us , this journey is suddenly disrupted. This disruption is unexpected, unwanted and uncalled for. Unwarranted . We are told that we have to step off the caravan of life , break journey at some unknown destination . A Station called Cancer. Of course we don't want to get off the train . We don't want to be separated from our co travellers . We do not want our life interrupted . But it is . No amount of Control Alt Delete is going to change things and it seems like there is no going back to that what now seems like an utopian state we inhabited . Washed ashore on some strange land we find ourselves alone , afraid , confused, angry and totally unwilling to accept this new stage in our life. We are in denial and keep asking of the universe , Why Me? Isolation , fear , uncertainty, awareness of our mortality is then overtaken by the practical needs the situation warrants. What is and where can we access treatment and how will we find the finances for the treatment. Of course it does not end with treatment for it is then that the healing will begin .

Gradually , these people who had to leave the caravan of life and find themselves alone and lost , unable to continue on the main track towards a common destination find the strength and courage to seek and build a new , parallel track in order to continue living , with hope and dignity . Their destination they realised had not changed . All they had to do was create a parallel track , a new normal .

This was possible when the realisation came that one was not actually alone and without resources . There were others in the same space , diagnosed and alone and eager to share their concerns and seek answers to the many queries they too had . And advocates at hand to guide and support them giving them a safe and secure platform upon which to share and learn from each other . Surviving cancer and living a normal fruitful life can be and is a reality .

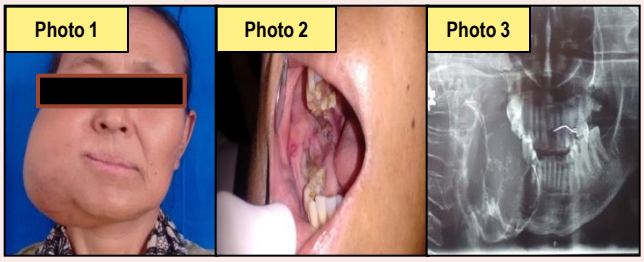
LATE PRESENTATION OF AMELOBLASTOMA: A Rare Case of Neglected Slow Growing Tumour

Dr. Y Indibor Singh
Dept. of Radiotherapy, RIMS, Imphal

Case History:
A 55 years old lady presented to our OPD with H/O slowly enlarging painless tumour on right lower jaw for last 9 years (photo-1 & 2). It has become painful during last 2 months. Many doctors had seen her and could not diagnosed properly. Repeated FNAC samples were inconclusive. She was investigated and found to have S/S of slow growing benign tumor syndrome. Oral examination showed ulcero proliferative growth affecting lower gum and alveolus. Externally a large 10X 12 cm swelling fixed to mandible and extending to Sub mandibular region was noted. Xray-Panorma mandible showed soft tissue swelling with soap bubble appearance (photo-3). CXR and USG abdomen WNL. CT scan Jaw and neck confirms an osteolytic lesion of right mandible. Teeths are loose but no dentigerous cyst detected. Cervical lymph nodes were enlarged involving level I ,II and III.

Diagnosis: Mandibular Ameloblastoma.

Treatment: Patient underwent Hemi mandibulectomy with LN dissection. And reconstructive surgery of mandible with fibular graft at RIMS. Resection margins were free.



Discussion:
Ameloblastoma is a rare benign tumour of Jaw usually affects mandible but maxilla can also be affected which is more aggressive type. They are a slow growing tumour type though benign in nature if left untreated they become massive and debilitating. 20 to 30 % will transform into malignant type and usually metastasize to lung. The diagnosis is missed by physicians who are not familiar with this disease and FNAC is often inconclusive. A dentigerous cyst may not be found however the clinical history and soap bubble appearance of mandible is diagnostic. This tumour arises from the epithelial rest cells in the mandible which form the enamel of teeth during development. Histologicaly the pallissading pattern of low columnar cells are pathognomonic. For treatment in early stage curettage is done but recurrence rate is high. A total clean resection is usually curative but in case of recurrence or resection margin not free local radiation therapy is given. However the role of RT is undefined and no effective chemotherapy is available. RT alone is given in painful massive unresectable tumour.

Conclusion:
A case of late presentation malignant ameloblastoma is presented due to physicians ignorance and for its rarity. A high index of suspicion is key to early diagnosis and proper curative early treatment. Even in this late stage curative surgical resection with reconstruction was performed and is successful.

EDITOR'S NOTE

With the blessing of all of the members the newsletter has now entered into the 3rd year. It is indeed a great pleasure that this year there is a great enthusiasm amongst the members for the article submission. I sincerely thank all the members of AONEI and people who are contributing for publishing the newsletter periodically.

Last year AONEI conducted academic meetings at Imphal (Annual Meeting) , Dibrugarh (Colo - Rectal cancer CME), Guwahati (Breast Cancer Update) with the participation of AONEI members and invited faculties from all over India. The new AONEI (www.aonei.in). website is functional and members are requested to make most use of it for sharing and exchanging ideas and knowledge.

Besides the academic activities AONEI is also doing Community and Social work as per its aims on regular basis. Members are actively getting involved in cancer awareness programmes.

This year the annual conference will be held at Silchar (Cachar Cancer Hospital & Research Centre) and we extend our gratitude towards the organizing committee for hosting the meeting. Hope to meet you all at Silchar.

Dr. Vikas Jagtap
Editor & Web In charge
drvikasj@yahoo.co.in
aonei_secretary@aonei.in

Correspondence:-
- aonei_secretary@aonei.in
- **Dr. Y Indibor Singh, Secretary, AONEI;** drindibor@yahoo.com
- **Dr. Vikas Jagtap, Editor and Website Incharge, AONEI;** drvikasj@yahoo.co.in
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RESPONSE EVALUATION CRITERIA IN SOLID TUMORS (VERSION 1.1)
RECIST: Imaging Basics

Dr Pratap Kakati MD (PGIMER)
Consultant Radiologist, Ayursundra One Stop Medical Centre, Guwahati

Response evaluation criteria in solid tumors are a set of public rules used to assess tumor burden in many anticancer drug trails in order to provide an objective assessment of treatment to therapy. They were initially introduced and published in 2000 and have undergone subsequent revision in 2009 **(RECIST 1.1).** Although improvements of clinical symptoms and survival rate are considered the ultimate proof of the effectiveness of therapy, primary endpoints based on radiological measurements are increasingly used to assess therapeutic effects. Such radiological measurements give early and objectively based information.
The criteria can be used with CT, MRI or conventional radiography if the lesion clearly visualized.
To understand the **RECIST** rules one need to know the some terminology i.e. Measurable vs. Non-measurable and Target vs. Non-target. Measurable lesions are the once which have at least one lesion that can be measured in at least one dimension using calipers. Application of unidirectional rather than the bidimensional measurements showed no difference in response and progression rate of disease process. From among the measurable lesions one can select a target lesion. Once a lesion is target it is always target even if falls below the size limits for what is considered measurable at baseline.
Measurable lesions must have a longest diameter of $\geq$ 10 mm on CT with a slice thickness of $\leq$ to 5 mm or $\geq$ 20 mm on non helical CT with a slice thickness of > 10 mm or a longest diameter of $\geq$ 20 mm on conventional radiography. Non-measurable lesion include small lesion with a longest diameter of < 10 mm, skeletal metastasis without soft tissue component, ascites, pleural effusion, pericardial effusion and leptomeningeal disease etc.

All measurable lesions up to a maximum of 2 lesions per organ and 10 lesions in total that are representative of all involved organs should be identified as target lesions. Target lesions should be selected on the basis of their size (lesions with the longest diameter) and their suitability for accurate repeated measurement. A sum of the longest diameter for all target lesions will be calculated and reported as the baseline sum longest diameter. The baseline sum longest diameter will be used as the reference by which to characterize the objective tumor response. All other lesions should be identified as non-target lesions and should also be recorded at baseline. Measurements of these lesions are not required, but presence or absence of each should be noted throughout follow up.

Lymph nodes merits special mention since their normal anatomical structures which may be visible by imaging even if not involved by the tumor. In lymph nodes the short axis rather than the long axis should be measured. In lymph node with short axis diameter < 10 mm regarded as normal, lymph nodes $\geq$ 10 mm but < 15 mm regarded as non target lesion and lymph node $\geq$ 15 mm regarded as target lesion. The assessment criteria are then used to essentially classified disease into four categories.

Evaluation of target lesions:
Complete response : Disappearance of all target lesions. Any pathological lymph nodes must have reduction in short axis to <10 mm.

Partial response : At least a 30 % decrease in the sum of diameters of target lesions taking as reference the baseline sum diameters.

Progressive disease : At least a 20 % increase sum of diameters of target lesions taking as smallest sum diameters. In addition to the relative increase of 20 % the sum must demonstrate an absolute increase of at least 5 mm. Appearance of one or more new lesions also considered as progressive disease.

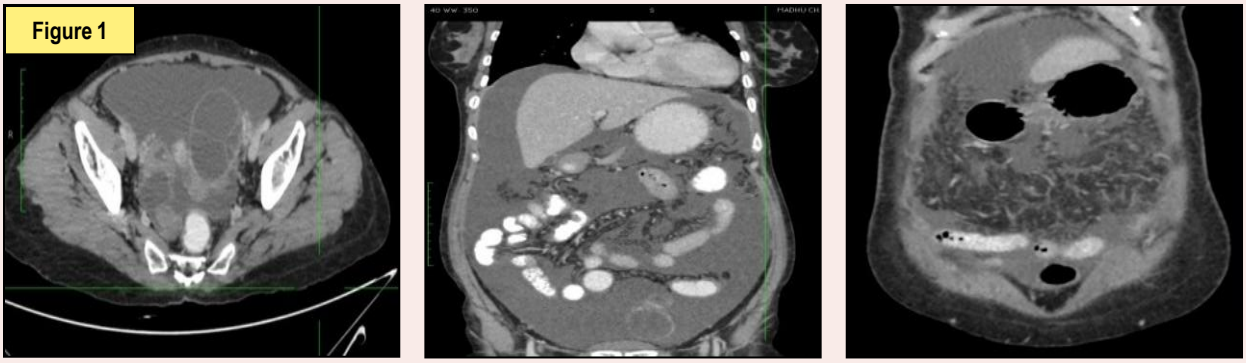
Stable disease : Neither sufficient shrinkage to qualify for partial response nor sufficient increase to qualify for progressive disease.

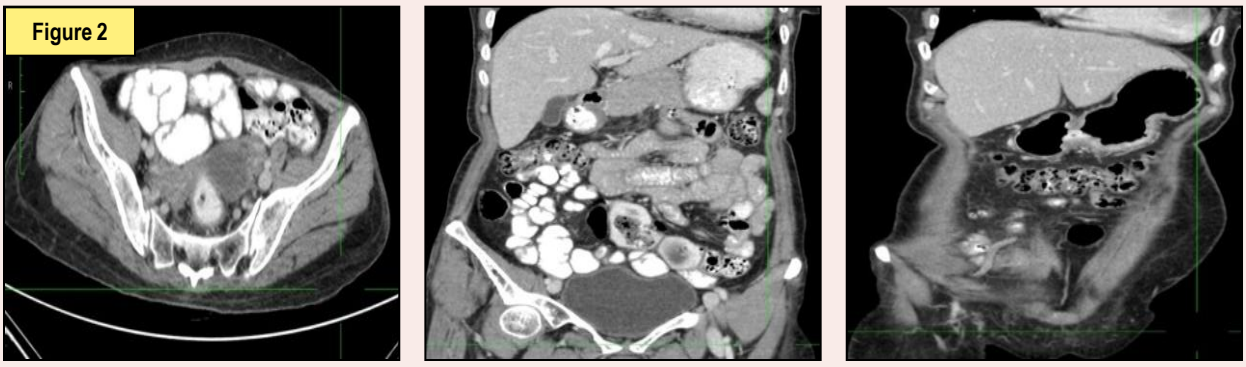
Evaluation of Non-target lesions:
Complete response : Disappearance of all non target lesions and normalization of the tumor marker level : All lymph nodes must be non pathological in size (< 10 mm in short axis measurements)
Stable disease : Persistent of 1 or more non target lesion and or maintenances of tumor marker level above the normal limits.
Progressive disease : Unequivocal progression of existing non target lesions. Appearance of one or more new lesions also considered progression.

RESPONSE ASSESSMENT (Example):

Figure 1: Baseline CT image of the pelvis shows bilateral ovarian tumors with ascites and omentoperitoneal thickening.

Figure 2 : Follow-up scan shows partial regression of the ovarian lesions (> 30%) with complete disappearance of the ascites and omentoperitoneal thickening. No new lesion was seen. Overall features would be partial response.





Conclusion: Familiarity with the revised RECIST guideline is essential in day to day oncologic imaging practice to provide objective tumor response. Although improvement of clinical symptoms and survival are considered the effectiveness of anticancer drugs. Radiological measurements are increasingly used to assess therapeutic effects, give early and objectively based information. It is expected functional imaging methods will be incorporated in the next RECIST update.

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AONEI Annual Conference (Imphal)

Cultural Program - AONEI Annual Conference (Imphal)

ONCOPLASTIC SURGERY- State Of Art In Breast Conservation

Dr. Poulome Mukherjee (Mch)
Surgical Oncologist, Cachar Cancer Center aand Research Institute, Silchar

Breast conservation surgery has been popularised over the last few decades as a viable alternative for mastectomy. The ever increasing evidences have proven the safety and long term oncologic outcomes equivalent to mastectomy. Studies have also shown a positive psychological effect over the patients and significant improvement of QOL in patients opting for breast conservation as against mastectomy.

During the initial phase of breast conservation the lumpectomy cavity was left for filling up with seroma. This technique provided a fair cosmetic outcome during the immediate post-operative period but the cosmesis deteriorated progressively over time after application of radiation.

Changing this idea was one of the hallmarks of oncoplastic breast surgery. The concept of superior and more durable breast cosmesis with preservation of oncologic safety and survival outcomes is the cornerstone of oncoplastic breast conservation surgeries.

The breast cavity after lumpectomy is filled with local glandular tissue from the breast or in case of larger resections, from the local area. Acheiving negative margins is the first objective which is confirmed by frozen section study. This is followed by mobilisation of breast tissue from the overlying skin and pectoralis muscle. Oncoplastic surgery is based upon integration of plastic surgery techniques for immediate breast re-shaping after wide excision of breast cancer.

ONCOPLASTIC PRINCIPLES: SELECTION CRITERIA

The three important elements to consider before deciding on the oncoplasty techniques:

1) Excision Volume- This is the most important factor for predicting surgical outcome and potential for breast deformity. Studies have suggested that more than 20% of reduction in breast volume after excision almost always results in deformity. The oncoplastic techniques therefore retain the breast shape in cases of larger volume excision by redistribution of breast tissue.

Oncoplastic procedures may involve redistribution or advancement of glandular breast tissue and in certain cases volume replacement by latissimus dorsi “mini flap”.

2) Tumor Location- lumps in the medial or lower inner quadrants are high risk zones for post resection deformity. The upper outer quadrant is a very favourable location and allows large resections without major distortion of anatomy.

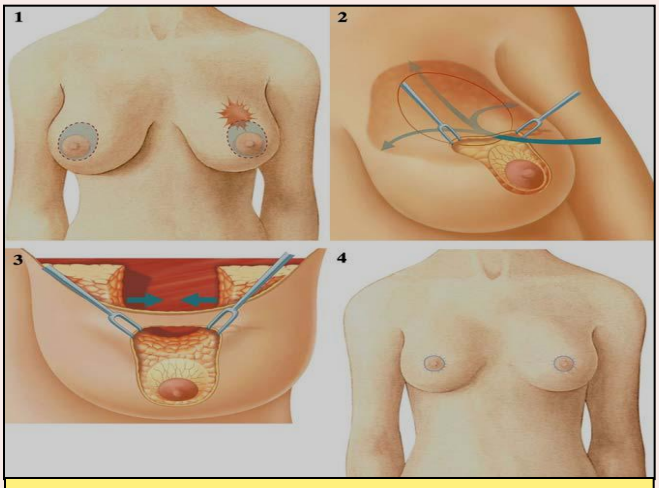
3) Glandular Density- The fat versus glandular composition of the breast is a factor to be determined before planning OPS procedures. The fat composition is usually graded on the Breast Imaging Reporting and Data System (BIRADS). The heavily dense breast (BIRADS 3/4) allows extensive undermining of gland for flap raising without causing fat necrosis as compared to sparsely dense (BIRADS 1/2).

BASIC SURGICAL STEPS FOR ONCOPLASTY

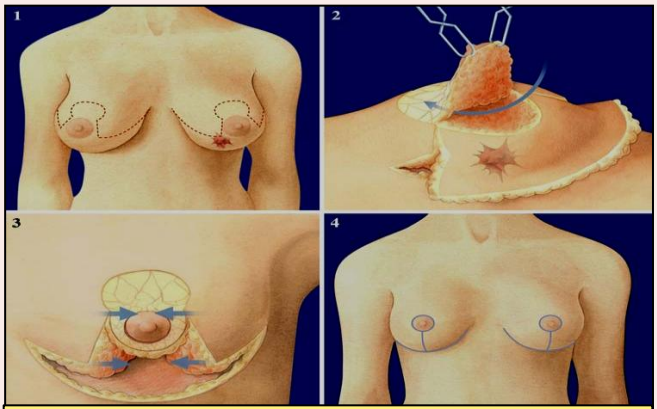
There are six basic steps for performing oncoplasty resections:

- Skin Incision
- Skin undermining or raising flap with optimal thickness
- NAC (nipple and areola complex) undermining
- Full thickness wide excision of the lump
- Glandular re-approximation which involves mobilising gland from underlying muscle and advancing towards the cavity.
- De-epithelialisation and NAC re-positioning which is important to prevent nipple deviation.

Besides the above surgical steps atlas for OPS procedures include step by step guide to OPS techniques customised for each quadrant of the breast. This article will mention some of the more commen locations. Detailed description of each procedure is beyond the scope of this article.



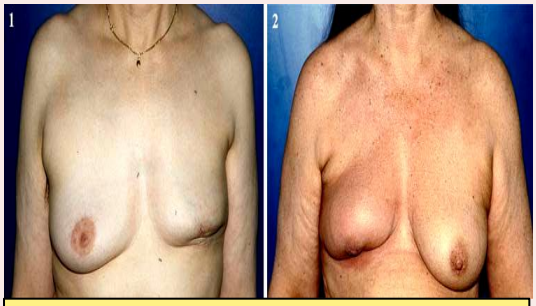
ROUND BLOCK MAMMOPLASTY TECHNIQUE



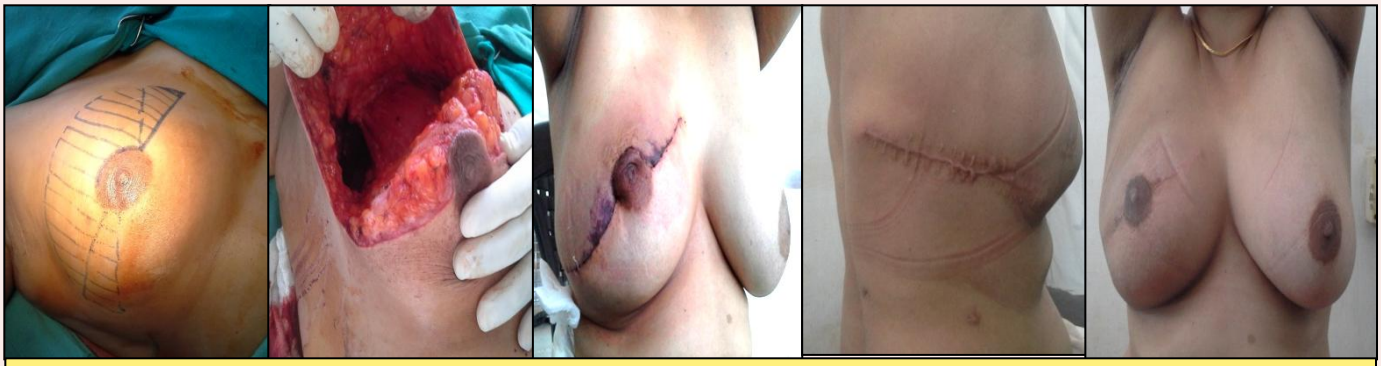
STEP BY STEP OF SUPERIOR PEDICLE MAMMOPLASTY FOR LOWER INNER QUADRANT TUMORS

ONCOPLASTIC SURGERY- STATE OF ART IN BREAST CONSERVATION

LOCATION OF TUMOR	PROCEDURE
Lower quadrant lesion (5-7o'clock)	Superior pedicle mammoplasty/inverted T
Lower inner quadrant (7-8o' clock)	Superior Pedicle Mammoplasty/ V scar
Upper Inner Quadrant (9-11o'clock)	Batwing
Upper quadrant (12o'clock)	Round block technique
Upper outer quadrant (1-2o'clock)	Racquet mammoplasty/radial scar
Outer quadrant (4-5o'clock)	Superior pedicle mammoplasty/ J scar
Lower outer quadrant or Central lesion	Grisotti's flap



THE TYPICAL BIRDS BEAK DEFORMITY AFTER LUMPECTOMY IN THE LOWER QUADRANT



ONCOPLASTY RECONSTRUCTION WITH LATISSIMUS DORSI MINI FLAP

APPLICATION OF IMUNOHISTOCHEMISTRY IN LYMPHOMA DIAGNOSIS: An Experience From A Under Resourced Laboratory In A Government Set Up

Gayatri Gogoi, MD

Assistant Professor of Pathology, Assam Medical College , Dibrugarh

COMMENTARY

New era begins with application of Immunohistochemistry method in the diagnosis and classification and treatment of lymphoma. Lymphoma is a highly heterogeneous disease and requires a multi –specialty approach. The cure rates of lymphoma are increasing and morbidities are decreasing with more active pharmacological agents and technological advancement in diagnosis. The Pathologists have a very active and crucial role to play in diagnosis treatment and prognosis . While pathologists must be equipped with modern methods and take primary responsibility for diagnosis and classification but involvement of clinicians is essential for successful outcome. While lineage is a defining feature of most ,in recent years there is greater appreciation of lineage plasticity within hematopoetic system. WHO emphasize the clinical knowledge both accurate diagnosis and definition in many cases such as MALT type, Nodal MZL/splenic, mediastinal large cell lymphoma vs DLBCL, most cases of T/Nk cells neoplasms.

Lymphoma diagnosis requires not only experience and skill but the laboratory set up loaded with huge numbers of primary antibodies available for immunohistochemisrty. These antibodies are costly which are directly proportional to the cost of the tests. In a public hospitals where rates of all services are subsidized and where 99% people are unable to afford the costs. So from where this amount would come to run the laboratory and for a hospital it is like having white elephant and leads to sustainability question . So the viability and sustainability of such testing facility is a big challenge. It requires courage and dedication to start such services knowing the limitations. But we did started it in spite of these challenges .With a limited panel of lymphoma markers initially in 2011 we experienced difficulties in reaching a technical stability with appropriate staining of slides. Again another difficulty was of sub-classification of low grade Non Hodgkin Lymphoma. For examples , without CD5 , it is impossible to differentiate Small lymphocytic lymphoma from marginal Zone lymphoma. So we did reported as Mature B cell lymphoma[low grade] and advised CD 5 study to assist the clinicians to begin the treatment in cases who can't go ahead to any cancer centre. Now a days there is a criteria to sub classify diffuse large B cell Lymphoma to Germinal centre type and Activated B cell type but due to cost of Mum 1 and BCL 6 , we used to report as only DLBCL with ki67 proliferation index. Precursor lymphoma such as B LBL and T LBL requires more B cell lineage markers besides CD20. T Cell NHL diagnosis needs again much more experiences to appropriately apply the various markers and we experience more extra nodal presentations.

Immunohistochemistry with limited markers lead to a diagnosis of broad category diagnosis such as peripheral T cell lymphoma in absence of Alk 1 or LMP 1 . Now Situation is much improved due to number of projects running in the laboratory so common reagents and antibodies are reducing the cost factor. Another important aspect of cost factor is availability of Ready to Use Antibody which are available in small quantity. Besides that RTU packs is greatly helping in quality assurance such as dilution in the procedure which is otherwise a big factor in discordance of staining and interpretation.

Incorporation of disease history, clinical diagnosis and radiological information to pathological report is a must in any lymphoma diagnosis .The histopathologic diagnosis is easier in most of the low grade B cell NHL as well as immunophenotypic profile. We have been using principles of small cell, intermediate size, large cells, low grade monotonous to high grade heterogenous appearance to subclassify and application of IHC accordingly . Pathologists should be more careful as chances of missing polymorphic and heterogenous NHL such as T cell NHL where neoplastic cells are scanty in a background of reactive cell population is very high. Our attempt was to classify it according to WHO classification . however when we fail due to various reasons then we didn't assigned specific subtype but kept in broad category. The factors of inability to classify was due to a]suboptimal morphology b)inexperience of rarer types morphology c) non-availability of some IHC marker d) inadequacy of tissue e) inconclusive expression profile in IHC.

However our clinical colleagues supported and cooperated with us with an optimistic feedback We have experienced limitations but that didn't not limit our learning, rather opened the door of the road to further leaning ...

UTILIZATION OF THE FOREHEAD-FLAP TO REPAIR SKIN DEFECT FOLLOWING MAXILLECTOMY: A Case Report

Dr. R.J.Das[1] , Dr. Kunal Ranjan[2], Dr. Ashok Kumar Das[3], Dr. Srimanta Kumar Medhi[1]

Assistant Professor Deptt of Head and Neck Oncology BBCI[1] ,Fellow Deptt of Head and Neck Oncology BBCI[2] Associate Professor Deptt of Head and Neck Oncology BBCI[3]

Abstract:

Local flaps have been commonly used in the reconstruction of facial defects left after excision of primary tumours. The forehead flap (median and laterally based) can be used to close defects in the cheek and floor of the mouth, its pedicle can be used to close defects in the maxilla. In a seventh century Indian medical document, the *Sushruta Samita,* describes a technique of using a flap from the forehead for nasal restoration. The forehead flap is relatively simple in concept and technique. It is widely used for nasal reconstruction. The importance of this flap is due to its versatility . This flap has been described as the most robust and dependable flap. In addition, it has the advantage of having large arc of rotation. Further, it provides good color matching at the host site, hair-free pedicle, and matching tissue texture. The severe arc of rotation usually does not compromise the blood supply, thus good vascularity is an additional benefit for wound healing. The flap is basically a paramedian flap and utilizes single supratrochlear/supraorbital **vessel**

Case Report:

A 51 year old male was admitted in the head and neck department of BBCI with chief complaint of swelling Right sided cheek for last 2 months. Proper clinical evaluation and investigations were done. The skin overlying the maxillary mass was fixed. CECT scan faciomaxillary region and biopsy was done from alveolar mass that came to be differentiated squamous cell carcinoma. The disease was staged as cT4aN0M0. CECT (fig 1) showed soft tissue mass eroding Left maxillary alveolus, hard palate and anterior portion of zygoma. Preoperative photographs were taken(fig 2 and fig 3). A modified lateral rhinotomy incision was made(fig 4) so as to include the skin defect and the left sided total maxillectomy with overlying skin excision with adequate margin was done. There was a (3 × 4 cm) defect not possible for local repair so laterally based forehead flap was used to repair with two staged procedures living behind controlled fistula as shown in the figures(fig 5 and fig 6). A controlled fistula was left over to get epithelised and was planned for closure later, so as to promote tension free wound healing . Soft tissue defect over forehead was covered wth SSG(split skin graft).. After a period of 3 weeks the forehead pedicle was detached and the fistula over the maxilla was also (figure7). Post operatively patient was put on adjuvant radiotherapy with concomitant chemotherapy.

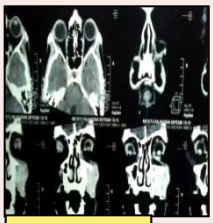


Figure 1

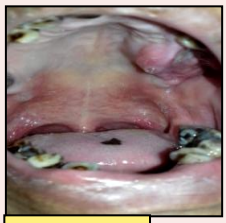


Figure 2

Figure 3

Discussion: Forehead flap was used by Sushruta in 600 BC for nasal reconstruction. Reliability of success of this flap is a major advantage which comes from the adequate blood supply and local availability of feeder vessels, i.e. supratrochlear/supraorbital vessels, and is a reason of its popularity[1]. This flap has been described as the most robust and dependable flap[2].


Figure 4


Figure 5


Figure 6


Figure 7

The primary blood supply is through supratrochlear vessels with multiple anastomoses to the dorsal, and supraorbital and angular arteries Usually the forehead flaps are about 5 cm in height from the eyebrow to the hairline; this measurement may be useful in estimating the tissue availability for reconstruction. It is used for reconstruction of defects which are more than 2 cm in diameter[3]. The flap is basically a two-stage procedure. Stage one involves marking for designing of flap, elevation, and insertion. Stage 2 involves the division of pedicle and reshaping of the tissues to achieve the normal anatomy of the area.

With all these advantages, there are two main limitations, viz., and the arc of rotation my compromise the blood supply of the flap. In a study done by M. Frans Noorman van der Dussen[4] the forehead flap can be used to close defects in the cheek and floor of the mouth, its pedicle can be used to close defects in the maxilla. This method was successfully applied in six patients. Less than 1% of our cases in the deptt of Head and Neck oncology are of Nose and PNS. The case on discussion is a rare presentation requiring repair of the large skin defect. Looking into the size of defect we thought forehead flap will be better choice.

IMAGING IN CERVICAL CANCER

Dr. Caleb Harris M.Ch. & D.N.B.(Surgical Oncology)
Surgical Oncologist, NEIGRIHMS, Shillong

Introduction

India accounts for over a fifth of the worldwide incidence of cervical cancer, which is the second most common cancer of Indian women(1). The survival rate varies from 80-93% for stage 1 to 15-16% for stage 4 as per data published in AJCC 2010 staging manual.

The predominantly followed staging system for cervical cancer is the Féderation Internationale de Gynécologie et d'Obstétrique (FIGO) staging system which was revised in 2009 after consultation with several other cancer societies, including the Americal Joint Committee on Cancer(AJCC). The FIGO staging relies only on the clinical features. While this enables the system to be adopted universally, there are shortcomings. Apart from the inter-observer variability, the lack of information on exact size of tumor(especially in endocervical lesions) and the lack of information on the nodal status is a major drawback.

Alvan Feinstein, in 1985, first used the term "Will Rogers Phenomenon" to describe the 'stage migration' observed in patients with cancer(2). This refers to the spurious improvements in stage- specific prognosis of patients, without the actual change in prognosis of individual patients. In cervical cancer, incorporation of information such as para-aortic nodal involvement may lead to stage migration. Though not necessary for staging, imaging is often performed for cervical cancer, even though the information is not incorporated into the FIGO staging system. This is to guide treatment planning.

With the easy availability of various imaging modalities, there is the dilemma over selecting the most appropriate tool. This article attempts to address this issue.

Need for Imaging

Prior to choosing a particular imaging modality, it is imperative to first ask oneself as to what is the question to which an answer is sought. In general, imaging is performed to assess the local extent- size of tumor and involvement of the parametrium and to assess the presence of nodal metastasis. Understanding the indications for adjuvant therapy following surgery for early cervical cancer will help us determine the appropriate imaging tool also. Table 1 shows the various risk categories into which patients are stratified following surgery for cervical cancer. Patients with any of the high risk factors will have to undergo multimodality therapy(surgery->chemoradiotherapy)(3), thereby increasing the morbidity and resulting in poor quality of life. Hence efforts should be made to identify these high risk factors prior to surgery, thereby avoiding surgery and administering chemoradiotherapy alone. While the size of the tumor, possibility of attaining clear margin and parametrial involvement can be assessed by clinical examination, it may not be possible in obese patients. Also, assessment may be incomplete in patients who are apprehensive. Though pelvic examination under anaesthesia may provide this information, imaging can offer an objective, non-invasive assessment.

Lymph node metastasis, shown in table 2, increases with the stage. In a study of patients who underwent surgery for FIGO Ia2 to IIb, parametrial involvement and tumor size > 2cm were independently associated with nodal metastasis(4). In general, imaging is not warranted in patients with small volume, early stage disease (Ib tumors and cervical tumor size less than 2cm). However, in larger tumors and when the size of the tumor cannot be ascertained well, as in endocervical tumors, imaging may be helpful.

In patients with locally advanced cervical cancer, who are generally treated with radiotherapy, evaluation of nodal involvement will help in planning the extent of radiotherapy fields.

MRI

Magnetic Resonance Imaging has better ability than CT scan to visualize the tumor and assess parametrial involvement, as it can delineate soft tissue structures well(5). It can also characterise the status of pelvic nodes. Therefore, MRI would be the modality of choice in early cervical cancers. However, the cost, non-availability, longer imaging duration and need for skilled radiologists are the limitations.

CT Scan

Computed Tomography scans are easily available, relatively inexpensive and can be easily interpreted by clinicians. Though its inferior to MRI in assessment of the parametrial involvement, it can help in studying the status of nodes, both pelvic and para-aortic. The latter is especially important in locally advanced cancers, where radiotherapy treatment planning would depend on this information.

FDG PET-CT

Functional imaging using Fluoro Deoxy Glucose is helpful in detecting nodal status and distant metastasis. It is not helpful in staging the local tumor. In early cervical cancers, the incidence of nodal disease is low and, when present, it is of low volume, which is generally below the resolution of PET-CT(6). However, it is relevant in locally advanced cervical cancers, wherein it can help in detection of para-aortic nodal disease, which may require extended field radiotherapy and also in detection of distant metastasis, which may change the intent of therapy(7). In India, the possibility of false positive nodal disease due to tuberculosis, can lead to dilemma in decision-making. Hence more Indian studies are needed before PET-CT can be incorporated routinely into practice.

PET-CT is recommended prior to extensive procedures such as pelvic exenteration, wherein detection of distant metastasis may preclude futile surgery.

Conclusion

The choice of appropriate imaging modality for cervical cancer should be guided by the clinical stage of the disease and the treatment plan. MRI is the best tool for assessment of the tumor extent, which is of relevance in early cervical cancers, while CT scan would be appropriate for locally advanced cervical cancers, wherein extent of nodal involvement needs to be ascertained.

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High Risk Factors **positive pelvic lymph node** **positive surgical margin** **parametrial involvement**	**Adjuvant Chemoradiotherapy (If any one of these high risk factors)**
Intermediate Risk factors **Lymphovascular space invasion(LVSI)** **>1/3rd stromal invasion** **Tumor Size> 4cm**	**Adjuvant Radiotherapy (If any two of these risk factors)**
Low risk **None of above**	**No Adjuvant Therapy**

Table 1

Stage	Pelvic LN (%)	Para Aortic LN(%)
Ia1	0.5	0
Ia2	4.8	<1
Ib	15.9	2.2
IIa	24.5	11
IIb	31.4	19
III	44.8	30
IVa	55	40

Table 2

DERMATOFIBROSARCOMA PROTUBERANS

Dr.Niju Pegu (Mch Trainee), Dr.B K Das, Dr. J Purkayastha, Dr. B B Borthakur, Dr. A Talukdar, Dr. P. Mandal, Dr. A Jain

Departement of Surgical Oncology,Dr. B Borooah Cancer Institue, Guwahati

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is an uncommon and locally aggresive mesenchymal neoplasm. DFSP constitutes approximately 1% of all sarcomas and <0.1% of all malignancies. Although DFSP may have been reported in the literature as early as 1890, Darier and Ferrand first described it in 1924 as a distinct cutaneous disease entity called progressive and recurring dermatofibroma. In 1925, Hoffman officially coined the term dermatofibrosarcoma protuberans. It is characterized by translocation of the *COL1A1 (chromosome 17)* and *PDG-FRB genes (chromosome22),* high rates of local recurrence and low risk of metastasis. This lesion typically presents in early or mid adult life body with no sex predilection. Most common site of involvement is trunk (50%), followed by extremities (30%) and head and neck (20%), but occurrence of DFSP in the mesentery is very rare and unusual. The cause of dermatofibrosarcoma protuberans (DFSP) is unknown. Laboratory studies have shown that chromosomal aberrations may contribute to the pathogenesis of DFSP; however, no evidence of hereditary or familial predisposition exists. In 10-20% of patients with this tumor, trauma at the site seems to be incriminated. The definitive diagnosis is established based on histological and immunohistochemical features. It stains positive for CD34. According to the literature, the overall risk for the development of metastatic disease is 5%, including 1% with regional lymph node metastasis and 4% with distant metastasis. The lungs are the most common site of distant metastasis. Usually, metastatic disease is preceded by multiple local recurrences.

HISTOLOGY

The cellular origin of DFSP is not clear. Evidence supports the cellular origin being fibroblastic, histiocytic, or neuroectodermal. DFSP manifests partial features of each. Therefore, many authorities suggest pluripotential progenitor cells, such as undifferentiated mesenchymal cells, may be the origin of DFSP, because they have the capacity to differentiate into all 3 cell types. It mainly consists of spindle cells in a fibrous stroma and variable amounts of collagen. Cultured DFSP tumor cells have increased growth in response to platelet-derived growth factor (PDGF)–beta.

MOLECULAR BIOLOGY

Cytogenetic studies reveal specific abnormalities in DFSP tumor cells, such as reciprocal translocations of chromosomes 17 and 22, t (17;22), and supernumerary ring chromosomes composed of interspersed sequences from bands 17(17q22) and 22(22q12). These rearrangements fuse the collagen type I alpha 1 (*COL1A1*) and the PDGF-beta chain (*PDGFB*, c-sis proto-oncogene) genes. The collagen promoter drives COL1A1 and PDGFB fusion protein production. The fusion protein is then processed into functional PDGF-B and subsequently interacts with the PDGF receptor on the cell surface of DFSP tumor cells. The activation of the PDGF receptor tyrosine kinase triggers the proliferation of DFSP tumor cells. Although cytogenetic testing of dermatofibrosarcoma protuberans (DFSP) is not established as a standard workup, reverse transcriptase polymerase chain reaction (RT-PCR) and fluorescence in situ hybridization (FISH) are suggested as screening tools for the presence of COL1A1-PDGFB fusion gene prior to initiation of oral imatinib molecular-targeted therapy.

CD34

It is a useful marker that allows differentiation of DFSP tumor cells from normal stroma cells and dermatofibroma. In dermatofibroma, tumor cells are positive for factor XIIIa and are rarely positive for CD34.

Immunostaining using CD34 as a marker is helpful in identifying tumor cells at the surgical margins, particularly when treating recurrent DFSP in which tumor cell fascicles are often interspersed with the scar tissue.

Imatinib as targated therapy in CD34 positive case

In recent studies neoadjuvant imatinib therapy for DFSP has been proposed in locally advanced or recurrent DFSP.

STAGING

The AJCC has not developed a staging system for DFSP. Because of its very low risk of metastasis, DFSP can be viewed as mostly a local disease. The following simple staging system published in “Short German guidelines: dermatofibrosarcoma protuberans” may be helpful in clinical use :

Stage I - Primary tumor, localized disease
Stage II - Lymph node metastasis
Stage III - Distal metastasis

TREATMENT

The gold standard treatment remains surgery with a gross margins of > 3 cm with or without radiotherapy. The surgery may be a wide surgical excision or Mohs technique. The later imparts a better outcome. The fundamental difference of these 2 techniques is the pathology processing. Usually, the specimen from wide excision is sectioned in conventional bread-loaf fashion, while the Mohs specimen is freshly frozen and sectioned en face along the margins. Mohs surgery requires less tissue removal and allows complete margin assessment. However, large tumor can be a challenge for this very time-consuming procedure.

DERMATOFIBROSARCOMA PROTUBERANS

TARGETED THERAPY

On October 19, 2006, the US FDA granted approval for imatinib mesylate (Gleevec) as a single agent for the treatment of DFSP. Imatinib mesylate is indicated for the treatment of adult patients with locally advanced, unresectable, recurrent, and/or metastatic DFSP. The recommended oral dose is 800 mg/d. Of note, fibrosarcomatous variants of DFSP lacking a genetic marker of translocation between chromosomes 17 and 22 may not respond to imatinib. The loss of the t (17, 22) cytogenetic marker in the fibrosarcomatous progression DFSP variant may represent progression of the malignancy.

Using imatinib as a preoperative therapy agent in locally advanced or recurrent DFSP may decrease tumor load, promote tumor cell apoptosis, and subsequently reduce the extent of surgery.

PROGNOSTIC INDICATORS

Age older than 50 years, regional lymph node involvement, extent of surgical excision, histologic features, high number of mitotic figures, increased cellularity, DNA aneuploidy, *TP53* gene overexpression, and the presence of fibrosarcomatous changes are poor prognostic indicators.

A CASE REPORT

A 62 years male with known case of recurrent DFSP was admitted with Lump Abdomen in our institute. He has past history of multiple excisions (3 times in right supraclavicular fossa and once laparotomy and excision of intraabdominal mass) and PORT. After admission he was evaluated with CECT abdomen and biopsy and immunohistochemistry. The reports show large mesenteric mass (fig.1) and spindle cell tumour (fig.2) with IHC CD34 positive (table.1).

Exploratory laparotomy was performed and large mesenteric mass (12x12cm) was found with adherent to jejunum at one point. En-bloc excision of mass with segment of jejunum was done. Final histopathological reports shows features of DFSP with minimal areas of necrosis and myxoid change and mass infiltrating the serosa and muscle layers of adjacent jejunum. Cut margins free. No post operative radiotherapy was given. Since then patient has been followed up without any evidence of reccurrence.

CONCLUSION

DFSP is a uncommon malignant sarcoma of skin but can develop anywhere including mesentery as a reccurrence. Clinical and morphological similar to other spindle cell lesions (eg. Fibrosarcoma, myxoid liposarcoma, leiomyosarcoma, fibrohistiocytoma, GIST, neurofibroma) may result in an inadequate diagnosis and treatment. This report highlighted the main clinical, morphological and immunohistochemitry for establishment of diagnosis of a recurrent DFSP in mesentery.

TAKE HOME MESSAGE

DFSP is a highly recurrent mesenchymal tumour and can occur anywhere in the body including mesentery.

UPCOMING EVENT

National Seminar on Diagnostic Pathology
4th-5th Feb 2017
Organised by: Department of Pathology
Dr. B Borooah Cancer Institute, Guwahati-16
Organizing Secretary: Dr. Anupam Sarma
dranupamsama@gmail.com, +91-9864016791

NE – AROI Zonal Chapter Annual Conference
October 2017
Aizawl (Mizoram)
Organizing Secretary: Dr. B Zothankima
+91-9436142185

Annual AONEI Conference
28th-29th Jan 2017
Cachar Cancer Hospital & Research Institute Sichar (Assam)
Organizing Secretary: Dr. Ritesh Tapkire
ritesh.tapkire@cacharcancerhospital.org,
+91-9854016164

Seminar: Head & Neck Cancer
May 2017
Organised by: Department of Head & neck Oncology &FHNO
Dr. B Borooah Cancer Institute, Guwahati-16
Contact:: Dr. A K Das, +91-99544139727

Regd. No. KAM/240/A-1/17 of 2006

AONEI : EVENTS AND ACTIVITIES

Cancer Awareness Program

Breast Cancer CME
Guwahati (Assam)

Colorectal Cancer CME
Dibrugarh (Assam)