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RESEARCH ARTICLE

PRIMARY CHONDROSARCOMA OF RIB: RARE NEOPLASM WITH ATYPICAL PRESENTATION

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Abstract

Background: Chondrosarcoma is uncommon malignant tumor of rib can have atypical presentation based on age, gender and clinical features. However, Computed Tomography is imaging modality of choice and helps aids in diagnosis and guiding treatment.

Case Presentation: This was a case of 84 year old lady, who presented as a breast lump. Core needle biopsy gave the differential diagnosis Malignant phylloides with heterologous cartilaginous component, Primary chondroid sarcoma, Metaplastic carcinoma. However, imaging findings led to diagnosis of chondrosarcoma arising from right 4th rib and hence patient underwent en bloc excision of anterior chest wall with chest wall reconstruction. Final histopathology was suggestive of grade 2 chondrosarcoma with medial margin microscopically positive. Patient was kept on 2 monthly follow up after surgery without giving any adjuvant therapy or additional surgery. She was disease free at 6 months follow up.

Conclusion: The incidence of rib Chondrosarcoma is rare and presents a diagnostic challenge. Surgical management with wide local excision and reconstruction remains the treatment of choice.

Introduction:-

Chondrosarcoma is common malignant tumor with most common site in the pelvis or long bones, but primary rib chondrosarcoma is rare with an incidence of 0.5 per million person-years.[1] However, chondrosarcoma represents the most common primary bone tumor involving the ribs and sternum .[2] It is included in the category of chondrogenic tumors, is designated as sarcoma arising from the cartilage matrix, and is associated with local destruction or metastasis. The main stay treatment is surgical resection, and a complete cure cannot be expected with chemotherapy or radiotherapy .[3]. Since the incidence of rib Chondrosarcoma is rare, we here present this case ,as it adds to the literature and give some insights regarding diagnostic challenge, atypical presentation and surgical management with reconstruction.

Case Report

84/F lady, resident of Ujjain Madhya Pradesh presented to the hospital with chief complaint of right Breast lump since 8 months which was progressive in nature, slowly increased in size with no associated complaint of pain, fever, axillary lump or nipple discharge. There were no complaints of anorexia, weight loss, cough, breathlessness, hemoptysis, headache, dizziness. She had no co-morbidities and no past surgical history. She had no addiction, post menopausal with normal bowel and bladder habits with adequate sleep and appetite.

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On general examination, she was averagely built and nourished with performance status of ECOG 1. She was haemodynamically stable with no signs of pallor, icterus or lymphadenopathy. On local examination, left breast and axilla was normal. A Single bony hard lump of size 8*7 cm was palpable in right breast , upper quadrant and upper inner quadrant and upper outer quadrant, with irregular lobulated surface, laterally extending till posterior axillary line.Lump was fixed to chest wall with skin involvement laterally.No peau d orange. Nipple areolar complex was pulled upwards with no involvement. No palpable right axillary lymphadenopathy or supraclavicular lymphadenopathy. Other systemic examination, examination of back and spine was normal with normal per rectal and per vaginal examination findings.

Patient had come with the outside incisional biopsy report of fibroadenoma with metaplastic calcification. Because of non availability of slides, slide review was not done. Imaging was done in the form of Magnetic resonance imaging of chest with cuts of Computed Tomography scan. It showed a large expansile lytic lesion of size 53*87*75 mm with associated lobulated soft tissue of altered signal intensity, hyperintense on T2, arising from right 4^{th} rib. It was heterogenously hyperintense with hypointense linear septations and bony spicules wihtin, reaching upto costochondral junction, infiltrating into overlying pectoral muscles and intercostal muscles with soft tissue extension into breast parenchyma, reaching upto skin laterally. On post contrast scan, heterogenous enhancement of soft tissue along septa and cyst walls with central necrosis seen with projection into the extrapleural thoracic cavity suggestive of neoplastic etiology of bony origin ? chondrosarcoma.

Core needle biopsy was done that showed fragmented cores of breast parenchyma with few tubules lined by bilayered benign epithelium. Focal small clusters of atypical cells were seen in stroma along with few tiny fragments of cartilage with variable nuclear atypia and multinuleation. Differential diagnosis was made : Malignant phylloides with heterologous cartilaginous component, Primary chondroid sarcoma, Metaplastic carcinoma.

Case was discussed in multidisciplinary board and with the clinic-radiologic diagnosis of chondrosarcoma, surgical resection was planned. Intrathoracic extent was first evaluated by thoracoscopy. Then en bloc excision was done with 2cm margins with overlying skin and underlying pleura along with 3^{rd} , 4^{th} and 5^{th} rib. Reconstruction was done using polypropylene mesh and latissimus dorsi flap reconstruction was done for skin and soft tissue cover. Duration of surgery was 4.5 hours and average blood loss was 300 ml. Patient was started mobilisation from post operative day 1 itself. In post operative period, patient developed sinus tachycardia which was managed with Ivabradine. On post operative day 5, patient developed left lower limb deep vein thrombosis, that was managed with unfractionated heparin followed by oral dabigatran 150mg twice a day. Pulmonary embolism was ruled out. Tachycardia settled in 2 days. Left lower limb edema was also reduced . Intercoastal drainage tube was removed on post operative day 9 and patient was discharged with stable vitals.

On final histopathology, microscopy was suggestive of cartilaginous matrix with lobules of cartilage, showing moderately increased cellularity with moderate atypia , occasional spindling and multinucleation of haphazardly arranged chondrocytes with mitosis of 8/10 hpf. No invasion into pleura was seen. No lymphovascular or perineural invasion was identified.

So, final Histopathology was: Chondrosarcoma, Grade 2, involving right 4^{th} rib, anterior soft tissue of breast, soft tissue posterior to 4^{th} and 5^{th} rib (extrapleural) with tumour size of 8.5*8.0*4.0 cm and no lymphovascular invasion. Gross margins were negative, however medial margin came out to be microscopically positive. Rest all margins were free .

Histopathology was discussed in multidisciplinary board decision was made to keep her on observation without any additional surgery or adjuvant treatment and to be on regular 2 monthly follow up after discussing with the patient and attendants. On 6 months follow up, patient was doing well with no locoregional or distant recurrence.

Discussion:-

Chondrosarcoma of rib is a very rare malignant tumour. Majority of patients present with painful progressive anterior chest wall tumour and its more common in females.[4] . CT scan is the imaging modality of choice. Intratumoral calcification on CT in the form of a “flocculent or popcorn” pattern has been described as the hallmark of Chondrosarcoma, that was also present in our case. and surgery in the form of wide local excision is the preferred treatment, irrespective of the tumor grade. Chest wall reconstruction is recommended for full-thickness defect of more than 5 cm or anterior resection involving more than three ribs.[5] Resection margins are classified into

curative, wide, marginal, and intralesional margins, according to the distance from the reactive zone around a tumor. As per the Surveillance, Epidemiology, and End Results (SEER) cancer registry database, tumor grade histologically is classified into four grades: well differentiated (grade 1), moderately differentiated (grade 2), poorly differentiated (grade 3), and undifferentiated (grade 4). Regarding the extent of the wide resection margin, the survival rate is significantly higher with a wide resection margin of approximately 2-4 cm. [6] Mayo group reported in 1985 where 10-years survival rate was 96.4% for those who had wide resection, compared with 65.4% for those who had local excision only. [7] In cases with microscopic positive margins, indications of additional resection are unclear. Data on cases with positive microscopic margins is limited and decision on reresection is to be taken on the basis of grade of lesion. [8] Small non randomized data is available on role of Doxurubin-based adjuvant therapy in grade 2 and 3 mesenchymal tumors. [9] The role of radiotherapy in high-grade tumors is also unclear and has failed to achieve better overall survival or cancer-specific survival. [10] Role of immune checkpoint inhibitors is also emerging.[11]

Rib chondrosarcoma has five-year mortality rate of 10%, a local recurrence rate of 17%, and a metastatic rate of 12%, relatively better outcome than general chondrosarcoma.[12] Tumor diameter greater than 5 cm, positive surgical margins, higher grade and local recurrence are poor prognostic factors.Long-term follow-up is essential, as studies have shown that 13% of grade 1 chondrosarcomas develop local recurrence and 1/3 of patients with local recurrence develop metastases [12] Post-operative surveillance consists of physical examination and thoracic imaging with chest X-rays or CT scans every six months for the first five years and then annually for a minimum of ten years.

The five-year survival rate for radical resection is reported to be 78% versus 15% for incomplete resection. The five-year survival rates are of 97%, 57%, and 39% for grades 1, 2, and 3, respectively.[13]

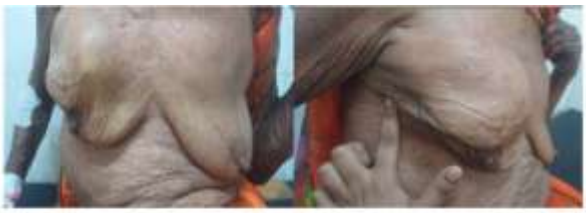

Figure 1. Preoperative pictures of right anterior chest wall mass

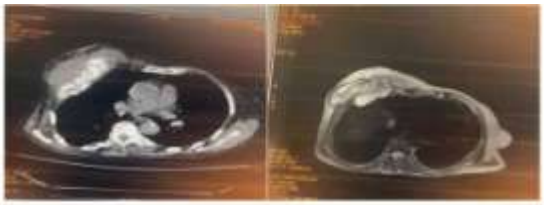

Figure 2. CT and MRI image of right anterior chest wall mass, arising from 4th rib

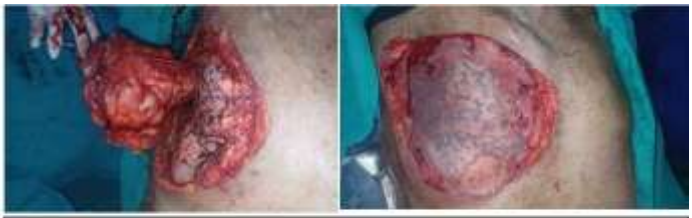

Figure 3. Intraoperative pictures

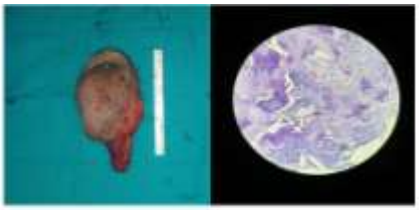

Figure 4. Excised specimen and Histopathological picture

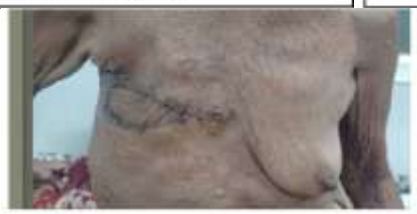

Figure 5. Postoperative clinical picture with Latissimus Dorsi flap

Conclusion:-

Primary Chondrosarcoma of rib is rare and can have atypical presentation. Diagnosis is based on CT imaging and surgical resection with reconstruction remains the main stay of treatment.

Declarations

Ethics approval and consent to participate: appropriate ethical approval and consent was taken

Consent for publication:

Consent to publish was taken.

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