VOLUME - 12, ISSUE - 07, JULY - 2023 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Pulmonary Medicine

A RARE CASE OF PNET OF CHEST WALL: A SKINS TUMOUR

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ABSTRACT Ewings sarcoma is second most common osseous malignancy in children and adoloscents. It may arise from extraosseous tissue of the chest wall as described in this case report of 24 year old who visited our tertiary centre. Also referred to as Askins tumour, this tumour is often missed in the initial stages. Patients in developing countries like India are often treated as cases of empema or tuberculosis leading to delayed presentations and further mismanaged. Approach to this condition requires involvement of multidisciplinary working groups with surgical management and chemotherapy being the main stay. Its association with poor prognosis is related to its local recurrence and distant metastasis.

KEYWORDS :

INTRODUCTION

Peripheral primitive neuroectodermal tumors (PNET) are a group of soft-tissue tumors of neural crest origin that arise outside the brain, spinal, and sympathetic nervous systems. An estimated 6.5% of primary lesions arise in the chest wall. These tumors occur primarily in children and adolescents (thirteen to sixteen years old) and are included in the differential diagnosis of malignant small round cell tumors, which include Ewing sarcoma, rhabdomyosarcoma, neuroblastoma and lymphoma.

CASE REPORT

24 year old female presented with complaints of left sided non radiating dull aching chest pain for 2 months with loss of appetite and loss of weight for 2 months of around 4 kg. No significant past or medical history present.

On examination, general condition was average with chest movements decreased on left side with signs of volume loss. Dull notes were percussed over the left hemithorax with decreased breath sounds felt on left side.

Persistent pain over chest wall forced the patient to seek medical help following which a chest xray done that revealed an opacity that occupied the entire left hemithorax. She was started on antitubercular treatment on clinical basis from a primary health facility following which there was no improvement and further referred to our facility.

On evaluation, blood reports were unremarkable. Usg thorax and abdomen showed mild pleural effusion with septation whereas bilateral subcentimetric lymph nodes were seen in usg neck which were round to oval lesion with central fatty hilum in submandibular region. Diagnostic tap of pleural fluid under usg guidance was performed. Pleural fluid was transudative with lymphocytic predominance and no malignant cells were seen on cytology.

CECT chest showed large poorly circumscribed heterogeneously enhancing soft tissue density in left pleural space compressing left lung segments medially. Lesion measured about 14.2 *14.6 *11.6cm approx. seen closely abutting mediastinal pleura and pericardium over the left ventricle. Few enlarged discrete necrotic and non necrotic mediastinal lymph nodes , larger node in pretracheal region measuring about 1.1*1 cm.

Usg guided fnac and biopsy revealed round cell tumour with ewings pnet being most probable diagnosis. Immunohistochemistry report showed positivity for CD99, synaptophysin and chromogranin. Negative for desmin, vimentin, EMA, LCA, TdT, Pan CK suggestive of Primitive neuroectodermal tumour.

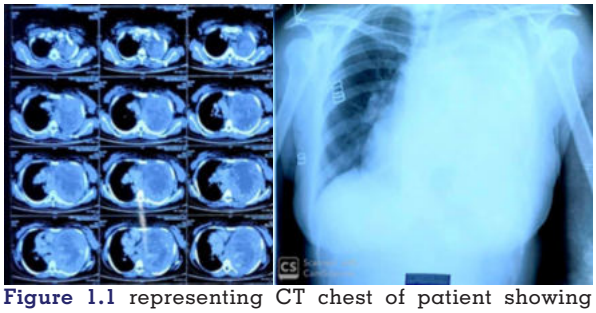


Figure 1.1 representing CT chest of patient showing heterogenous mass occupying left hemothorax with minimal pleural effusion. The picture on side representing Chest Xray of the same patient with opacity in left lung field.

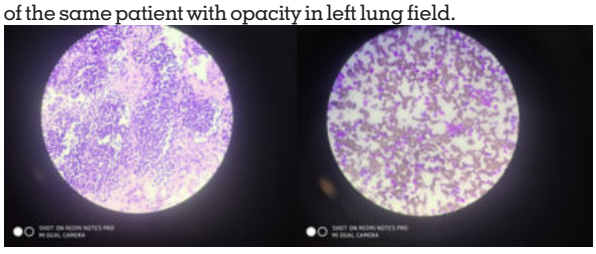
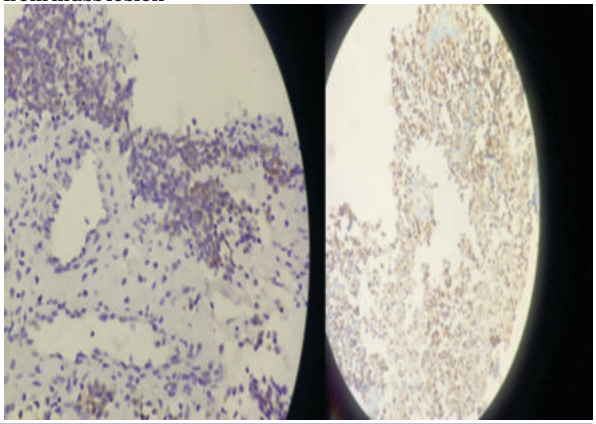


Fig 1.2. round cell tumour cells visualized on fnac and biopsy from mass lesion



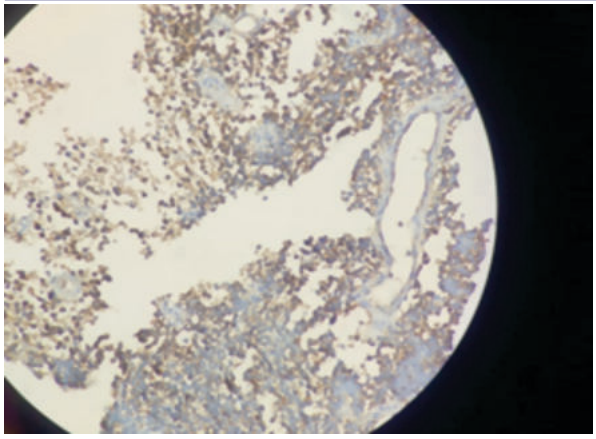


Fig 1.3 Immunohistochemical Staining Revealing Positivity For CD99, synaptophysin and chromograninA

DISCUSSION

The Ewing sarcoma family of tumors (ESFT), characterized histologically by primitive small round cells of neuroectodermal origin, includes classic osseous Ewing sarcoma, PNET, Askin tumor (Ewing sarcoma of the chest wall) and extraosseous (soft-tissue) Ewing sarcoma(1). Extraskeletal Ewing sarcoma represents a small subset of the ESFT, though the exact incidence has not yet been ascertained (2). While more frequent sites of ESFT include the retroperitoneum, paravertebral space, and chest wall (3), specific organs of involvement described in the literature also include the kidneys, pancreas, colon, uterus, and ovaries (1). The prognosis for treated patients has been reported to be similar to that for Ewing sarcoma patients (4). The lung represents a rare organ of primary involvement. Initially reported in 1989 (5),it occurs in a wide age distribution spanning the pediatric and adult age range, with the mean age of onset around 20 years as compared to 15 years for osseous Ewing (3), and occurring more commonly in males than in females. There is high probability of missed diagnosis, specially at primary and secondary level health care practitioners, due to rarity of the disease. This is further compounded by nonspecific clinical presentation, overlapping with other common diseases. In our case study, the predominant symptoms were chest pain with loss of appetite and weight for a duration of 2 months.

Painful mass in the chest was one of the common clinical presentation, which often may be associated with fever and malaise [5]. Cough with fever and hemoptysis also were reportedas common clinical presentations reported by previous case studies [3]. Shortness of breath and chest pain also were reported as other clinical presentations [1,6].

EES family of tumors present with typical imaging characteristics. Primary pulmonary Ewing sarcoma usually presents as a circumscribed solitary mass with heterogenous appearance both on noncontrast and on contrast-enhanced CT [7]. In our case as well, a heterogeneously enhancing mass was described with no bony involvement on CT . Bone marrow aspiration could not be done due to issues in consent for confirmation.

Proliferation of small round cells with scanty and clear cytoplasm, round to oval nuclei, finely granular chromatin, and inconspicuous nucleoli are the typical histological feature. Strong reactivity to CD99/MIC-2 and vimentin is another vital feature [8]. Markers of neural differentiation like S-100 and neuron-specific enolase may also be present in few cases [9]. Cytokeratin positivity was reported in about one-fifth of the cases. Other supporting diagnostic features include, translocation t(11,22)(q24;q12) in fluorescent in situ hybridization (FISH) and reverse transcription-polymerase chain reaction (RT-PCR) [10].

Histological examination of a sample taken from our patient revealed markedly cellular tumor composed of malignant round cells stained positive for CD99 and negative for TTF1, LCA and pan-cytokeratin. Hence, both morphological and immunohistochemistry findings were consistent with the diagnosis of EES.

Reciprocal chromosomal translocation for t(11;22) (q24;q12) could not be done due to limited financial resources .

Multimodality treatment, consisting of surgical resection followed by chemotherapy with or radiotherapy, is needed for effective management of EES. Ifosfamide, etoposide, cyclophosphamide, vincristine, doxorubicin, carboplatin, and actinomycin have been documented to be effective in management of EES [11].Pazopanib was reported to provide a long-term progression free survival by a recent study.

CONCLUSION

Clinicians should consider primary extraskeletal Ewing's sarcoma as one of the differential diagnosis, when encountered with a young patient presenting with large mass with no obvious evidence of primary extra thoracic disease. Immunohistochemistry plays a vital role in definitive diagnosis. Multidisciplinary team approach is important in timely diagnosis and effective management to prevent serious adverse consequences.

Compliance With Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Funding None.

Ethical approval Not necessary.

Human and animal rights statement No animals were involved in this study.

Informed consent Informed written consent was obtained from the patients to use their clinical details for publication in a de-identified manner.

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