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RESEARCH ARTICLE

PULMONARY EWING SARCOMA/PRIMITIVE NEUROECTODERMAL TUMOR : A CASE REPORT

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Abstract

Extraosseous Ewings sarcoma is an extremely rare neuroectodermal tumor,Although thoracic Ewing sarcoma/primitive neuroectodermal tumor (PNET) usually developson the chest wall, there have been reports of primary Ewing sarcoma/PNET of the lung Wereport the case of a 30-year-old manwho presented with right-sided pulmonary mass.Radiology, histopathology, and immunohistochemistry confirmed the diagnosis of primarypulmonary Ewings sarcoma, the patient wastreated by chemotherapy (regimen ofvincristine, doxorubicin, cyclophosphamide alternating with ifosfamide, etoposide) andradiotherapy.

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Introduction:-

The Ewing's sarcoma family includes several malignant cancers that typically develop from bone. James Ewing, an American pathologist, originally characterized it in 1921. Ewing sarcoma (EWS) and primitive neuroectodermal tumors (PNETs) are members of the same family of malignant . The chest wall, pelvis, and limbs are typical sites for EWS PNETs (1). It primarily affects the ribs when it is in the thorax (Askin tumor). Although metastases can have an impact on the lungs, primary lung involvement is relatively uncommon. We describe a primary extraskeletal Ewing's sarcoma of the lung that was discovered and treated at our facility in this report

Case description :

A 33-year-old chronic smoker presented with cervicobrachial neuralgia persisting for a year, Unresponsive to analgesic treatment, and concurrent with a weight loss of 10 kg over three months. Clinical examination revealed signs consistent with Claude Bernard Horner syndrome.
Radiological assessment unveiled a sizable tumor, 13 x 12 x 13 cm in size, occupying the upper lobe of the lung, with invasion into the mediastinal pleura, contralateral pulmonary nodules, and right axillary adenopathy (Figure 1).

A CT-guided biopsy of the lung lesion was performed, and the histological and immunohistochemical aspects favored a proliferation of round cell tumors, initially indicating a tumor from the PNET (Peripheral Primitive Neuroectodermal Tumor) group.

A molecular study using FISH was conducted to detect the rearrangement of the EWS gene and translocation (X, 18), revealing 100% of hybridized cells and 60% of positive cells. The baseline bone scintigraphy did not reveal any secondary bone locations

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Following a multidisciplinary team discussion, the patient was deemed unsuitable for surgery. They were initiated on chemotherapy using the VDC-IE protocol along with GCSF support. (vincristine, doxorubicin, cyclophosphamide, ifosfamide, etoposide).

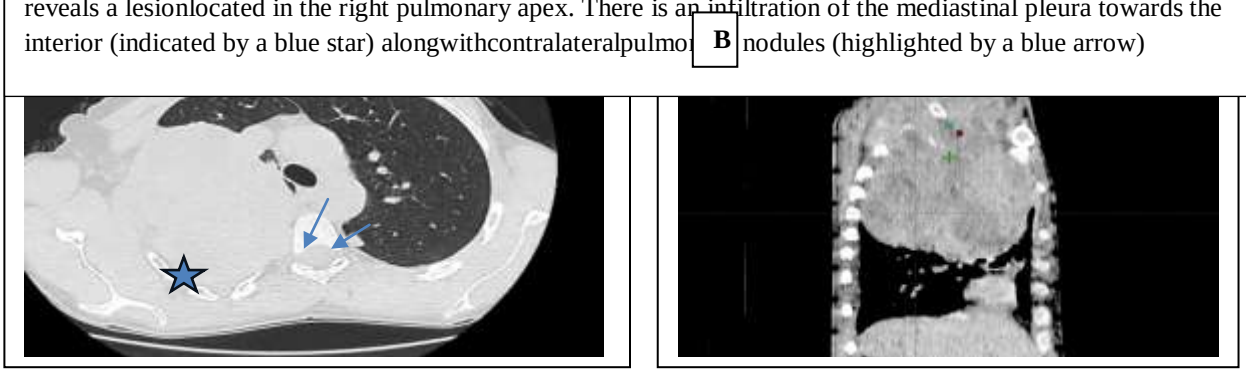
After three cycles of chemotherapy, the radiological assessment showed varied responses: progression of the pulmonary tumor and axillary adenopathies, alongside stability in metastatic lesions (Figure 2).

External radiotherapy consisting of 30 Gy administered in 10 fractions, followed by a 20 Gy boost in 10 fractions (Figure 3), resulting in decreased tumor size and central necrosis (Figure 4).

Following the initial treatment, involving three cycles of VAI-type consolidation chemotherapy (vincristine, doxorubicin, ifosfamide), ongoing clinical and radiological progression led to the decision to initiate second-line chemotherapy. This involved adopting the TEMIRI protocol, employing temozolomide and irinotecan.

As the patient's condition advanced further, necessitating a transition to second-line chemotherapy, the treatment plan shifted to the TEMIRI protocol. This protocol involved administering temozolomide at 100 mg/m²/day from day 1 to day 5 and irinotecan at 20 mg/m²/day for consecutive days, aligning day 1 with Day 21 of the treatment cycle

Figure 1 : A thoracic CT scan in axial view (pulmonarywindow - A) and sagittal view (mediastinalwindow - B) reveals a lesionlocated in the right pulmonary apex. There is an infiltration of the mediastinal pleura towards the interior (indicated by a blue star) alongwithcontralateralpulmonary nodules (highlighted by a blue arrow)



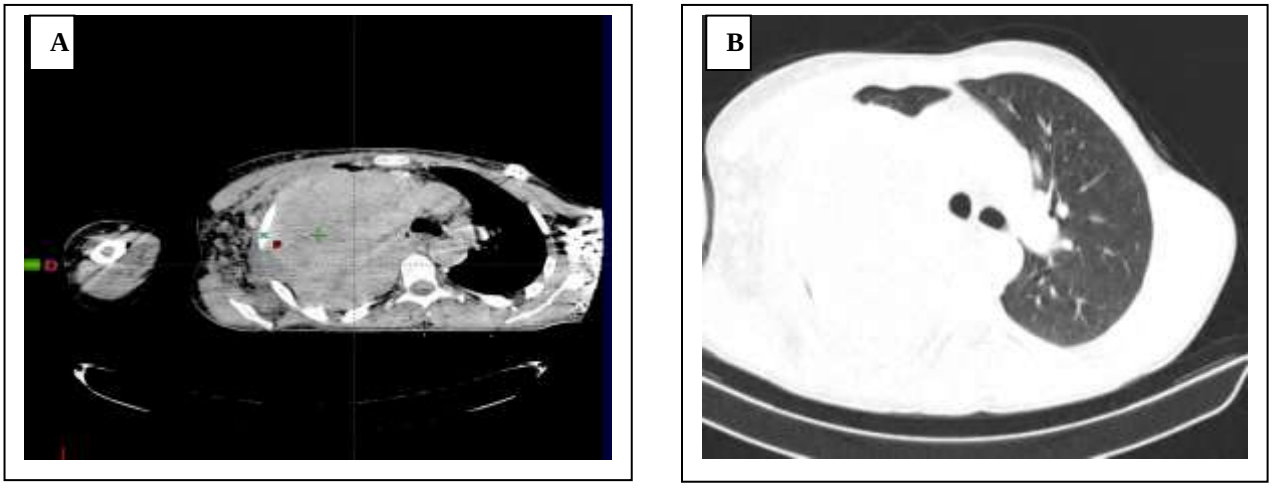


Figure 2 : Afterthree rounds of chemotherapy, a thoracic CT scan evaluationdemonstratestumor progression in theleftupperlung area, accompanied by the emergence of lymphadenopathy in the ipsilateralaxillaryregion

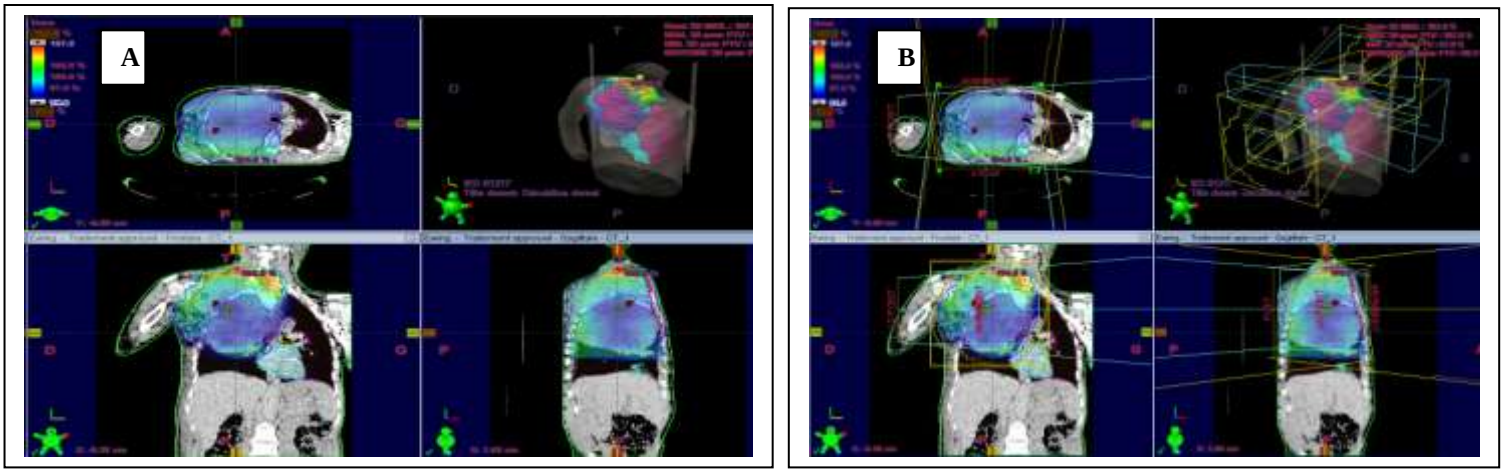


Figure 3 : Series of axial, sagittal, and coronal scans from the dosimetric CT depict the distribution of the dosage within the PTV (marked in blue) (A), alongside the configuration of beamsacross the three planes (axial, coronal, and sagittal) (B)

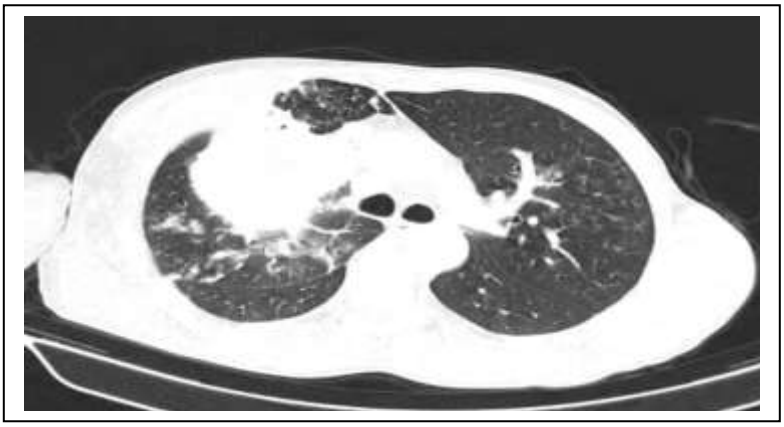


Figure 4 : Thoracicevaluation CT scan 3months afterradiotherapyshowedobjectivelyobservedregression in bothtumor size and central necrosis.

Discussion:-

Ewing sarcoma ranks second in prevalence among primary bone cancers, with only osteosarcoma being more common (2-3). Typical sites for primary extraosseous tumors include the buttocks, paravertebral muscles, chest wall, and retroperitoneal area. Instances of primary ES of the lung are rare and scarcely documented in the literature (4-6). The initial identification of PPES dates back to 1989 by Hammar et al (7).

Due to the rarity of primary pulmonary PNETs, thorough clinical and radiographic examinations are necessary to distinguish between metastatic tumors originating from an extrapulmonary primary site.

EES affects individuals aged 2 to 30, peaking around 20, with a slight male predominance. Symptoms vary depending on the location, typically causing discomfort and swelling of nearby structures due to the mass effect.

Clinical details are lacking, and symptoms of primary pulmonary disease often relate to discomfort and swelling of adjacent structures due to the tumor mass.

CT scans aid in determining tumor size and confirming the entirely extraosseous nature of soft tissue

masses. Histology predominantly confirms the diagnosis of EES.

The translocation t (11, 22) (q24; q12) characterizes Ewing sarcoma, present in 85% of patients, leading to the development of the EWS-FLI1 fusion gene (8).

Given its rarity, there are no standardized guidelines for managing EES. Treatment typically involves a multimodal approach, combining surgical resection with radiotherapy or chemotherapy to effectively manage the condition.

The prognosis depends on obtaining disease-free surgical margins and the extent of anatomical dissemination to adjacent structures like bone, pleura, and the epidural space (9).

First-line chemotherapy typically includes vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide. While pazopanib has shown efficacy in pulmonary Ewing's sarcoma cases, it's not considered a conventional treatment (10-11).
Radiotherapy may be considered for individuals without surgical options, those with unresectable tumors post-induction chemotherapy, or those with removed tumors but positive margins.

Primary EFT of the lung is exceptionally rare, with only 17 documented cases in the literature (12-13-14). These cases were treated variably, using combinations of surgical resection and chemotherapy, sometimes with radiotherapy.

Insufficient current information hinders definitive conclusions regarding optimal treatment for primary EFT of the lung. However, studies suggest better outcomes with surgical resection for primary extraosseous Ewing sarcoma patients (15). Among patients followed up for two years, six out of eleven survived, whereas two out of five patients who passed away within two years had only undergone surgical care (14).

Conclusion:-

When a young patient presents with a massive lung mass without signs of primary extrathoracic disease, primary EES of the lung should be considered in the differential diagnosis. Immunohistochemistry is crucial for making a firm diagnosis in this case. For early detection and effective therapy of such a tumor, a multidisciplinary team must be included, hence the importance of expert centers for the management of sarcoma.

Competing interests:
None.

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