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Case Report

Oncology Section

Primary Ewings Sarcoma of the Lung

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ABSTRACT

Extraosseous ewings sarcoma is an extremely rare neuroectodermal tumour. We report the case of a 30-year-old female who presented with right sided pulmonary mass. Radiology, histopathology and immunohistochemistry confirmed the diagnosis of primary pulmonary Ewings sarcoma. This case highlights the fact that Ewings sarcoma should be considered in differential diagnosis of patients presenting with pulmonary mass.

Keywords: Extraosseous, Ewings sarcoma, Lung

CASE REPORT

A 30-year-old female was admitted with complaints of right sided chest pain, shortness of breath and cough with scanty white expectoration since three months. Respiratory system examination was suggestive of right sided massive pleural effusion with shift of mediastinum to the left. Chest radiograph PA and lateral view [Table/Fig-1] were suggestive of right sided pleural effusion. Computed tomography (CT) of the chest and abdomen [Table/Fig-2] revealed a large, relatively well-defined, moderately heterogeneously enhancing mass lesion involving right hemithorax with pleural deposits, bilateral axillary nodes and gross right pleural effusion.

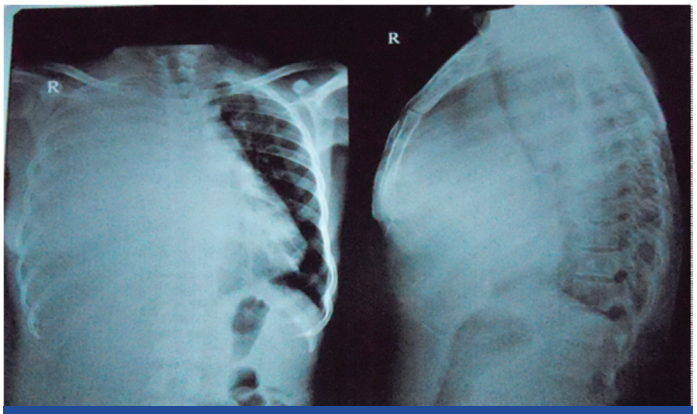
The patient underwent CT guided needle biopsy, which revealed small round cells with scanty cytoplasm, round to oval nuclei, fine granular to vesicular chromatin suggestive of malignant small round cell tumour. Immunohistochemical staining for pancytokeratin and CD 45 were negative. The tumour was MIC-2 positive. Thus, the histological and immunohistochemical findings were compatible with Ewings sarcoma [Table/Fig-3a-d]. The patient thereafter underwent PET scan, which did not reveal any evidence of an occult primary. Thus, a definitive diagnosis of primary Ewing sarcoma of the lung with nodal metastasis was made. She was started on chemotherapy but unfortunately the patient died after 15 d.

DISCUSSION

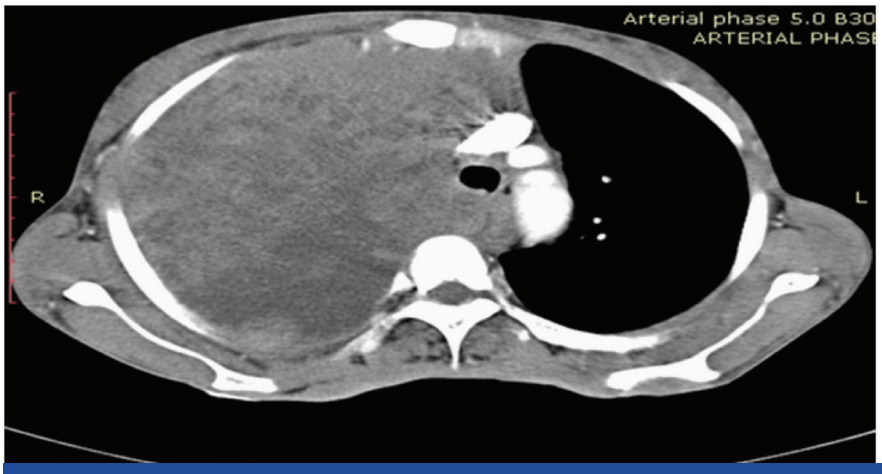
Ewings sarcomas are relatively rare neuroectodermal tumours that primarily arise from the bone [1]. Extraosseous ewings sarcomas have been reported but are extremely rare. It was first described in 1921 by James Ewing as an endothelioma of bone [2]. They are neuroectodermal tumours, which primarily arise in the bones and are the second most common primary bone tumour [1]. Translocation t (11, 22) (q24; q12) is pathognomonic of Ewing sarcoma, occurs in 85% of patients and it gives rise to the formation of the EWS-FLI 1 fusion gene [3].

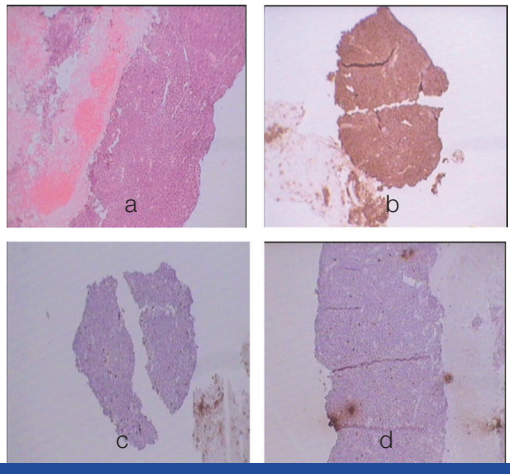
Extraosseous ewings sarcoma is extremely rare. We reviewed the literature using the search terms “Primary Ewings sarcoma lung” and found that only 16 cases have been reported so far. The first case was reported by Hammer et al., [4]. As per previous case reports, the patients were in the age group of 4- 67 y and 10 of the 16 cases were males [Table/Fig-4].

Histologically, the tumour consists of a proliferation of small round cells with scanty and clear cytoplasm, round to oval nuclei, finely granular chromatin, and inconspicuous nucleoli. It is Periodic acid schiff positive due to the presence of cytoplasmic



[Table/Fig-1]: Radiograph of the chest PA view showing massive right sided pleural effusion





[Table/Fig-2]: Computed tomography of the chest showing large, relatively well-defined, moderately heterogeneously enhancing mass lesion involving right hemithorax
[Table/Fig-3]: a) Ewings sarcoma as monotonous sheet of small round cells (Hematoxylin and eosin, original magnification 10×). b) Tumour immunoreactive for Mic-2. c] Tumour negative for CD45. d] Tumour negative for Pancytokeratin

Author	Year	Age	Sex	Presentation	Treatment	Follow up	Remarks
Hammer et al., [4]	1989	64	Male	-	Chemo+ Surg + Radiotherapy	-	-
Catalan et al., [5]	1997	29	Male	-	Chemo+Surgery	-	-
Tsuji et al., [6]	1998	25	Female	Intrapulmonary mass	Surgery	Death after 2y	No evidence of extrapulmonary involvement by the tumour at presentation
Tsuji et al., [6]	1998	15	Male	Intrapulmonary mass	Chemo+Surgery	No evidence of recurrence for 2 y	No evidence of extrapulmonary involvement by the tumour at presentation.
Imamura et al., [7]	2000	41	Male	Tumour in the left upper lung	Chemo+Surgery	No recurrence after 22 mnth	-
Imamura et al., [7]	2000	30	Female	Tumour in the right lower lung	Chemo+Surgery	No recurrence after 16 mnth	-
Kahn et al., [8]	2001	18	Male	Right middle lobe mass	Surgery (Right Middle lobectomy)	Death after 2y	2 years after surgery, there was local recurrence for which patient underwent right upper and lower lobectomy
Mikami et al., [9]	2001	17	Female	Right lower lobe mass	Chemo +Surg + Radiotherapy	Death after 3mts	Metastases in mediastinum and right thoracic wall detected 3 months after surgery
Takahashi et al., [10]	2006	8	Male	Right upper	Chemo+Surgery	No recurrence for 9 mts after surgery	-
Young Lee et al., [11]	2007	67	Male	Left lower lobe mass	Surgery +Chemo	-	No evidence of metastases at presentation
Antelo et al., [12]	2009	22	Female	Right lower zone mass	Chemotherapy	-	No evidence of metastases at presentation
Hancorn et al., [13]	2010	44	Male	Right upper lobe mass	Surgery	-	Was found to have cerebral metastases 5 weeks following surgery
Siddiqui et al., [14]	2011	15	Female	Mass involving entire right hemithorax	Chemotherapy + radiotherapy	Died after 8 mnth	Metastatic nodule in the left lower lobe
Ichiki et al., [15]	2012	42	Male	Right lower lobe mass	Surgery + chemotherapy	No recur ence for 6 mnth after surgery	-
Alsit et al., [16]	2013	4	Female	Left upper Zone mass	Surgery + chemotherapy	-	-
Andrei et al., [17]	2013	31	Male	Mass in the lingual of left lung	Surgery +Chemotherapy + Local radiationtherapy	-	-
Present case	-	30	Female	Mass involving entire right hemithorax	Chemotherapy	Died	-

[Table/Fig-4]: Cases of primary pulmonary ewings sarcoma reported so far

glycogen. Histologic differential diagnoses include small cell carcinoma, malignant lymphoma, alveolar rhabdomyosarcoma, neuroblastoma. Tumours have a strong reactivity to CD99/MIC-2 and vimentin. In some cases, they may be positive for markers of neural differentiation like S-100, neuron specific enolase and 20% of cases are positive for cytokeratins. Demonstration of translocation t (11, 22) (q24; q12) by fluorescent insitu hybridisation (FISH) and/or reverse transcription-polymerase chain reaction (RT-PCR) is used to support the diagnosis [1].

Due to its rarity, there are no specific guidelines for the treatment of this disease. The treatment should be aggressive and should consist of surgery followed by chemotherapy and radiotherapy.

CONCLUSION

We have described an extremely rare case of primary pulmonary ewings sarcoma. Though rare, it should be considered in the differential diagnosis of children and adults presenting with primary pulmonary mass.

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