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...
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 neuroendocrine lung neoplasms.

**CONCLUSION**

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

**REFERENCES**


[7] Alsit et al., [16] 2013 4 Female Left upper Zone mass Surgery + chemotherapy - - -

[8] Andrei et al., [17] 2013 31 Male Mass in the lingual of right lower lobe Surgery + Chemotherapy + Local radiationtherapy - - -

[9] Present case - 30 Female Mass involving entire right hemithorax Chemotherapy Died - -

**Table/Fig-6:** Cases of primary pulmonary ewing's sarcoma reported so far
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