



### Case Report

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## Unusual case of adult onset Askin's Tumour – Case report and review

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### Abstract

Askin's tumour is a small round cell neoplasm of primitive neuroectodermal origin in the thoracopulmonary region. This tumour is predominantly seen in childhood and adolescence and is extremely rare in adults. We are reporting an unusual presentation of this tumour in a 35 year old woman with massive pleural effusion. The atypical manifestations in our case were clinical picture of massive pleural effusion with ipsilateral pleural mass and sparing of the thoracic cage, negative pleural fluid cytology and pleural biopsy negative and the presence of pulmonary nodules.

**Keywords:** Askin's tumor, Massive pleural effusion, Monomorphous round cells.

### Introduction

Askin's tumour is a small round cell malignancy of primitive neuroectodermal origin in the thoracopulmonary region. In the original account by Askin and Rosai, this tumour was described as a rare clinicopathological entity of childhood and adolescence with an extremely poor prognosis<sup>[1]</sup>. In this case report, we are describing this childhood tumour presenting in a thirty five year old woman, with unusual features.

*Key message:* Askin's tumor should be considered as one of the causes of malignant pleural effusion in the middle aged woman.

### Case Report

A 35 year old previously healthy woman presented with progressively worsening breathlessness, cough and chest pain of one month duration. She had significant weight loss during the same period. There was no history of fever, hemoptysis, contact with tuberculosis or recent travel. She was a homemaker and was not exposed to toxins or chemicals. General examination showed a woman with medium build and anemia. Respiratory examination revealed tracheal deviation to the right, dull note on percussion and markedly diminished breath sounds on the left side. There was no lymphadenopathy, chest or breast mass.

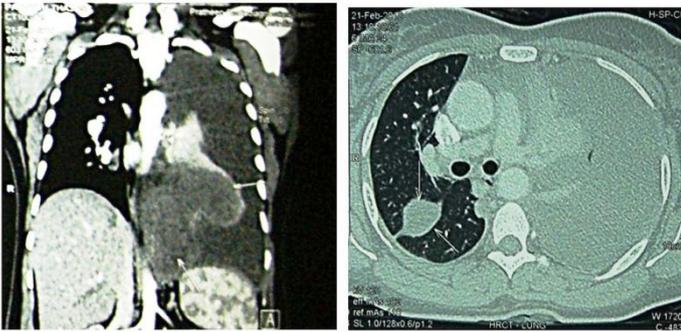
Her hemoglobin was 8.4 g/dl and the total and differential count were normal. Chest radiograph was suggestive of left pleural effusion with mediastinal shift. A diagnostic thoracentesis was performed and haemorrhagic fluid was obtained. The protein content of the fluid was 5.2 g/L and the cell count of 800 cells per mm<sup>3</sup> suggesting that she had exudative pleural effusion. The cellular response in the pleural fluid was predominantly neutrophilic. Pleural fluid adenosine deaminase was negative. Interestingly, cytological examination of the pleural fluid was negative. Because of worsening of breathlessness and rapidly accumulating pleural effusion, intercostal tube drainage was done and about 600 ml of the fluid was drained daily. Contrast enhanced Computed Tomography showed pleural effusion and pleural mass in left lower hemithorax and few small nodules in the right lung (Figure 1). The thoracic cage including the ribs was spared. Mediastinal lymph nodes were not involved. Pleural biopsy was performed, which was normal. Repeated cytology of the pleural fluid for malignant cells was negative. After a week of inpatient supportive treatment (while an open biopsy of mass was being planned), the patient worsened abruptly and succumbed to her illness. Post-mortem biopsy of the mass showed monomorphous round cells with stippled (dispersed) chromatin, high mitotic activity and focal areas of rosette formation. Immunohistochemistry showed CD 99 positivity (Figure 2 & 3).

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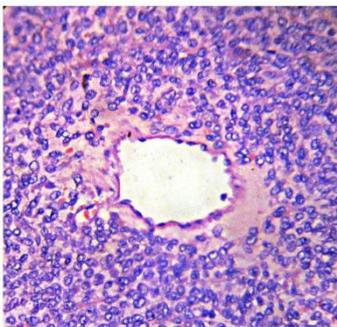
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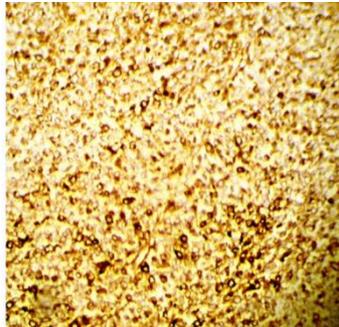
Cytokeratin (CK), epithelial membrane antigen (EMA) and Desmin staining were negative. These features were consistent with a diagnosis of Askin's tumour.



**Figure 1:** Chest CT showing massive pleural effusion with atelectasia in the left lung, pleural base mass in the left hemithorax, with sparing of the adjacent ribs and pulmonary nodule in the right lung.



**Figure 2:** Biopsy of the mass showed monomorphic round cells with stippled (dispersed) chromatin and high mitotic activity (haematoxylin & eosin x 40).



**Figure 3:** Biopsy of the mass showed diffuse CD 99 positivity in the malignant tumor cells

## Discussion

Small cell neoplasm is a rare entity in the thoracopulmonary region and includes pulmonary lymphoma, poorly differentiated adenocarcinoma, poorly differentiated squamous cell carcinoma and primary neuroectodermal tumour of the thoracopulmonary region (Askin's tumor). The differentiation of these tumors rest on their morphological appearance and immunohistochemical tests. Unlike other small cell neoplasms, the cells in Askin's tumour have nuclei with dispersed chromatin and one or more small nucleoli and express CD 99, an immunohistochemical marker.<sup>[2]</sup>

Askin's tumour was first described in 1979 as a rare, malignant small cell tumor arising in the soft tissues of the chest wall, occasionally in bone, or, rarely, in the periphery of the lung in children and adolescents. It is an undifferentiated sarcoma probably developing from embryonal migrating cells of the neural crest and is recognized as a primitive neuroectodermal tumor (PNET). This tumor chiefly occurs in girls during childhood and is characterized by restricted survival with the median survival of eight months. Research has shown that this tumour is characterized by reciprocal t (11, 22) (q24, q12) translocation<sup>[3]</sup>. Besides Askin's tumour, Ewing's Sarcoma and extraosseous Ewing's sarcoma share this chromosomal translocation and are classified as ES family tumors (ESFT). These tumours are only differentiated by their different anatomical locations.

The clinical presentation of Askin's tumour is nonspecific. The most common symptom is chest pain. Other symptoms in decreasing order of frequency are chest wall mass, breathlessness and fever. The

cardinal sign at presentation is chest wall mass or deformity. Pleural effusion is a common accompanying finding in Askin's tumour. Rib involvement is present in two-fifths of cases. It rarely involves pulmonary parenchyma and the common site of origin is the intercostal nerves. Bone, bone marrow and the lung are frequent sites of metastasis. Radiologic abnormalities range from unilateral chest wall mass, pleural mass, direct invasion to adjacent pulmonary parenchyma, pulmonary nodules to mediastinal lymphadenopathy<sup>[4]</sup>. Differential diagnoses of Askin's tumour include neuroblastoma, rhabdomyosarcoma and lymphoma. Establishment of the diagnosis is difficult and is made by the collective appraisal of the clinical, histologic and immunohistologic findings. Pleural fluid cytology has been reported as a useful tool for the diagnosis. Treatment includes radical surgical resection, neoadjuvant and adjuvant chemotherapy, and irradiation. Though, this aggressive tumour is highly chemoresponsive and radioresponsive, local recurrences and metastasis are often seen which accounts for the poor prognosis and short survival<sup>[5]</sup>. Other poor prognostic factors are age more than 26 years and presence of extraosseous tumour<sup>[6]</sup>.

Our case had several atypical features, in comparison to cases published in the literature. In the current case, the tumor occurred at thirty five years unlike the classical description as a childhood tumour. Contrary to the classical presentation as a chest wall mass, the presenting feature in this woman was massive pleural effusion with ipsilateral pleural mass and sparing of the thoracic cage. This is a very rare presentation of Askin's tumor. Interestingly, pulmonary nodules were noted in both lungs, a feature that has been previously has been reported<sup>[7]</sup>. Pleural fluid cytology, a useful investigation according to the published literature has been negative in our case. Even the pleural biopsy, which universally shows tumour invasion, was negative.

## Conclusion

To conclude, our case highlights an unusual cause of massive pleural effusion in a 35 year old woman, i.e. Askin's tumour. Apart from age, the atypical features of our case are massive pleural effusion with ipsilateral pleural mass and sparing of the thoracic cage; negative pleural fluid cytology and pleural biopsy; and the presence of pulmonary nodules. Though rare, Askin's tumor should be considered as a cause of malignant pleural effusion in the middle aged woman

**Conflict of Interest:** No conflict of interest noted. No financial implication.

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