CASE REPORT

Primary Pleural Epitheliod Hemangioendothelioma with Lung Involvement

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Abstract

The authors report a case of 50-year man who presented with 2-year history of dry cough, right sided chest pain, and shortness of breath. Chest X-ray revealed right-sided pleural effusion and left-sided opacity. Pleuroscopic pleural biopsy confirmed the diagnosis of primary pleural epitheliod hemangioendothelioma (EHE) with peripheral lung parenchymal invasion. Chest drain was inserted; and significant amount of fluid was drained, but lung failed to expand after 72 hours. Patient was planned for video assisted thoracoscopy (VATS) and also discussed with oncology department for chemotherapy; but he refused any further treatment, and left home against medical advice with chest drain in place. EHE originating from pleura is extremely rare with an aggressive clinical course and poor prognosis. To our knowledge, this is the first reported case of an EHE originating from pleura in South Asia and highlights the heterogeneous geographic distribution of tumor and demonstrates the need for a more systemic approach to all patients with unilateral pleural effusion.

Key Words: *Epitheliod hemangioendothelioma. Pleuroscopy pleural biopsy.*

INTRODUCTION

Epitheliod Hemangioendothelioma (EHE) is a very rare vascular neoplasm of endothelial origin with clinical behavior intermediate between angiosarcoma and hemangioma.[1] It is difficult to diagnose and most often presents as an incidental finding in young asymptomatic women. The most frequent presentation is bilateral multiple nodules. The tumor can simultaneously affect multiple organs, such as lung, liver, bone, soft tissue, skin, gastrointestinal tract, brain, mediastinum and spleen. It has a heterogeneous radiologic pattern. The natural history of EHE is highly variable and prognosis is very unpredictable, with life expectancy ranging from 1 to 15 years.[1] Pleural-based EHEs have been less frequently reported than those from other body sites.

CASE REPORT

A 50-year male patient presented to chest clinic with 2-year history of shortness of breath, dry cough, and right-sided chest pain. There was no history of fever, weight loss or night sweats. The symptoms worsened rapidly in 6 months prior to presentation; and pain was 7/10 on severity scale and sharp in nature. He was non-smoker with no prior medical or surgical history and denied any exposure to pets, birds or asbestos. Family history was unremarkable. He was a farmer by occupation. Chest examination revealed dullness and decreased breath sound on right side of chest while there was no clubbing, anemia, or lymphadenopathy. Base line investigations including complete blood count, urea, creatinine, electrolytes, liver function tests, and bleeding coagulation profile were all within normal limits.

Chest imaging including X-ray (Figure 1) and CT chest revealed right sided pleural effusion. Multiple, patchy, nodular infiltrates involving both upper lobes were also identified in CT chest. Workup for tuberculosis and lung malignancy was negative. Pleuroscopy showed hemorrhagic pleural effusion and multiple adhesions involving right side of pleura. Pleural biopsy report (Figure 2) showed evidence of EHE; fragments of fibrous tissue exhibiting an infiltrating neoplasm composed of

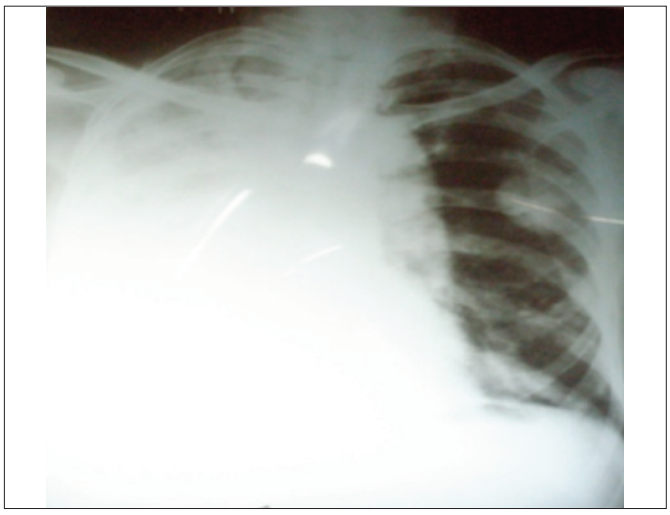

Figure 1: Chest X-ray PA view showing right sided pleural effusion and left sided rounded opacity.

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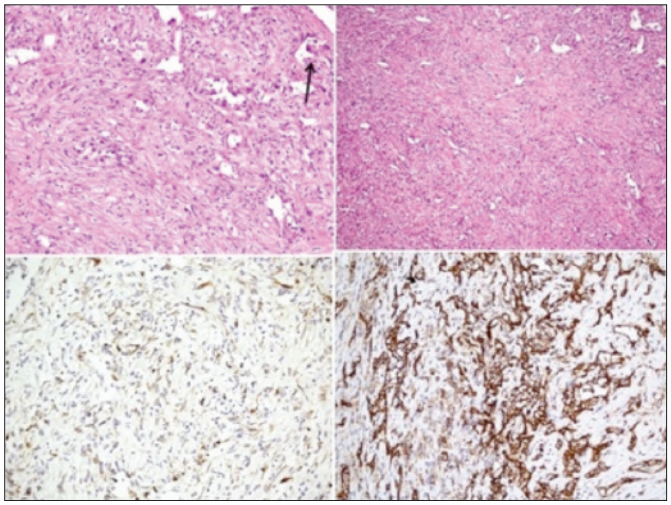

Figure 2: Biopsy slides of patient. Small interconnecting vascular channels seen lined by spindle shaped neoplastic cells. Imuno-histochemical stains demonstrated that the cells were positive for CD31, CD34, Cytokeratin Cm 5.2, and focal positive for cytokeratin AE1, AE3.

aggregates and small clusters of tumor cells. Small interconnecting vascular channels were seen lined by spindle shaped neoplastic cells. Immuno-histochemical stains were positive for CD31, CD34, Cytokeratin Cm 5.2, and focal positive for Cytokeratin AE1, AE3, confirming the diagnosis of EHE. Despite chest drain insertion and removal of significant amount of fluid, we were subsequently unable to re-expand the lung completely. Cardiothoracic opinion for VATS was planned. EHE usually is multifocal, so CT abdomen, brain, and bone scan were performed; but no evidence of metastasis was found.

Patient was diagnosed as primary pleural EHE with peripheral lung parenchymal invasion. Case was discussed with the oncology department and plan for chemotherapy was made; but he refused for any further intervention and left against medical advice.

DISCUSSION

EHE originating from the pleura is extremely rare. Till date, a total of 27 cases of pleural EHE have been reported in literature.[3] Only 4 cases had secondary lung involvement with primary pleural EHE.

Pleural EHE differs from its pulmonary counterpart (PEH) in many aspects. Pleural EHE mainly affects symptomatic older adult males with highly aggressive course of disease and a poor prognosis,[2,3] while PEH usually affects young to middle-aged asymptomatic females and usually progresses slowly with intermediate behavior.[4]

The poor prognostic factors of PEH include the presence of respiratory symptoms, pleural effusion on chest radiograph, extensive intravascular, endobronchial and interstitial tumor spread, hepatic metastases, peripheral lymphadenopathy and the presence of spindle cells in the tumor.[5]

Differentiation of pleural EHE from PEH, lung malignancy with pleural metastasis and malignant mesothelioma has important prognostic and treatment implications. The difference can be made on the basis of radiological, histological and imuno-histochemical grounds.[6] Anti-vimentin antibody, MNF 116, D2-40, anti-CD31, and anti-CD34 antibodies have definite role in making diagnosis.[3]

Regarding therapeutic options, there are no clear guidelines. Successful trial of surgical excision of the nodules has been reported in symptomatic patients, while no therapy in asymptomatic individuals is considered. However, because of extensive involvement, complete surgical resection of tumor is usually not possible and no other effective treatment has yet been established.[2] Different radiotherapy regimens and chemotherapeutic agents have been tried in the past but no clear benefit has been observed.[6] Most commonly used agents were carboplatin and etoposide.[3]

To the best of authors' knowledge, this is the first reported case of primary pleural EHE from South Asia. Cases from Korea, China, Europe, and America have been reported and our case highlights the heterogeneous geographic distribution of tumor and demonstrates the need for a more systemic approach to all patients, especially elderly group with unilateral pleural effusion, to exclude all malignant causes.

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