

Alveolar haemorrhage with pleural effusion as a manifestation of epithelioid haemangioendothelioma

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ABSTRACT: A 22 year old male was admitted with haemoptysis. A chest X-ray showed bilateral confluent alveolar infiltrates. Bronchoscopy revealed blood oozing from all bronchopulmonary segments. Open lung biopsy disclosed bilateral effusions and large necrotizing nodules with pleural adhesions. Histological examination showed tumour cells, which were negative to epithelial and embryogenic markers but positive to factor VIII. This confirmed the diagnosis of an epithelioid haemangioendothelioma. This rare tumour, usually has an indolent course, whereas in our case it was complicated by alveolar and intrapleural bleeding.

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Epithelioid haemangioendothelioma (EH) is a rare malignant tumour originating from the pulmonary small vessels. It maintains the endothelial marker, the factor VIII-associated protein. In addition to the lung, it has been reported to grow in the liver, mediastinum and diaphragm [1, 2]. Clinically, EH has been considered an indolent, non-aggressive tumour that displaces pulmonary parenchyma over a number of years by slowly enlarging tumour nodules [3]. We describe a case with a very rare presentation, diffuse alveolar haemorrhage and aggressive clinical course.

Case report

A 22 yr old man was admitted with haemoptysis, mild dyspnoea and anaemia of 10 g·dl⁻¹. His past history was unremarkable, with the exception, 6 months previously, of unexplained chest pain with a normal chest film. He had no fevers, chills, night sweats, chest pains, arthralgias or weight loss.

He appeared pale and was mildly tachypnoeic. Nose and throat examination did not disclose the site of bleeding. Auscultation of the lungs revealed bilateral diffuse crackles. Cardiac and abdominal examination were normal. There was no digital clubbing or cyanosis. Blood clotting was normal and no occult blood was found in the stools.

The patient's chest film (fig. 1) demonstrated bilateral confluent alveolar filling, mainly in the lung bases, and extending into the right wall with obliteration of its costophrenic sinus. Fibreoptic bronchoscopy on admission revealed blood oozing from all bronchopulmonary segments. No endobronchial lesions

were identified. Cytology showed the presence of haemosiderin-laden macrophages but no tumour cells. Cultures for bacteria, mycobacteria and fungi were negative. Subsequently, a chest computed tomography (CT) scan (fig. 2) revealed bilateral pleural effusion and large intrapulmonary nodular infiltrates.

An open lung biopsy was performed by sternal thoracotomy. One and a half litres of bloody fluid was drained from each pleural cavity. The parenchyma showed large 5×5 cm necrotizing nodules and pleural adhesions.

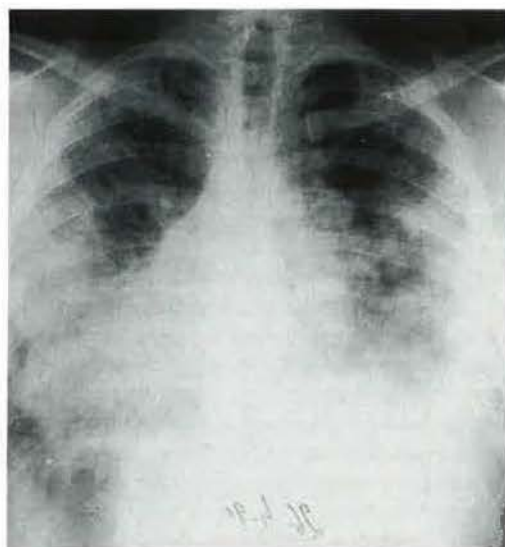


Fig. 1. - Posteroanterior chest X-ray shows diffuse bilateral alveolar infiltrates extending to the right wall with obliteration of the costophrenic sinus.

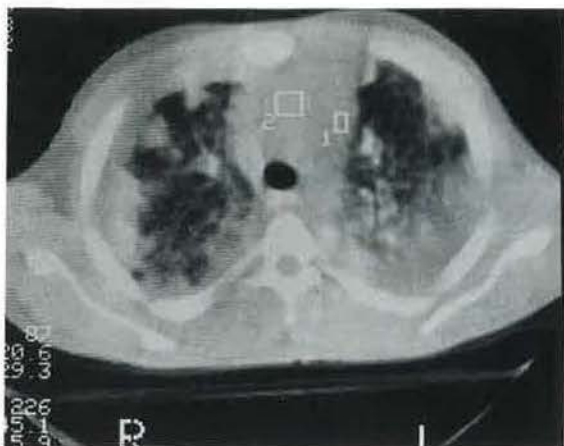


Fig. 2. - Computed tomography (CT) scan of the chest shows multiple large irregular nodules in both lungs, with pleural infiltrates.

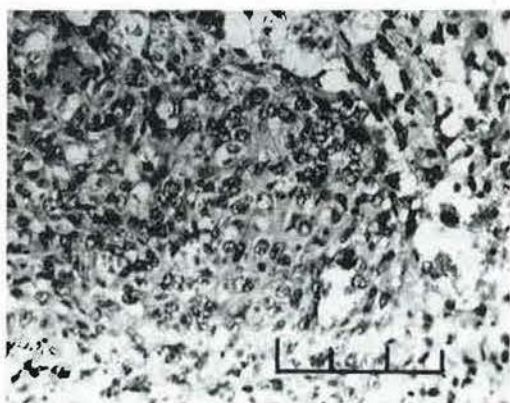


Fig. 3. - Haematoxylin and eosin stain of an open lung biopsy (magnification between bars 1×800). Papillary neoplastic growth consisting of both tumour cells and hyalinizing sclerosis.

Histology revealed the presence of polygonal, monomorphic and occasional rare atypical mitoses (fig. 3). In some areas, papillary neoplastic growth was seen originating from capillary walls and protruding into vessel lumina. Hyalinizing sclerosis was visible with thickening of the capillary basement membrane, alveolar haemorrhage, and haemosiderin-laden alveolar macrophages. Periodic acid-Schiff and mucin stains were negative. Stains for factor VIII were positive, whereas those for keratin and epithelial antigen membrane were negative. Antibodies to carcinoembryonic antigen and to S-100 protein were non-reactive.

The condition of the patient progressively deteriorated. He was overwhelmed by opportunistic infection and died two months after admission.

Discussion

We have presented a rare malignant tumour, which usually progresses slowly, but in our case had an aggressive course. The presentation with diffuse alveolar haemorrhage and pleural effusion is unusual. BEVELAQUA *et al.* [4] have described pleural effusions as a very rare manifestation of this tumour, whereas CARTER *et al.* [5] presented a single case of EH with alveolar haemorrhage.

Initially this tumour was called "intravascular bronchioalveolar tumour". Later, due to its endothelial origin, it became epithelioid haemangioendothelioma [2, 3, 6, 7]. Clinically and histologically, the tumour can be distinguished from haemangioma and conventional sarcomas. Unlike ordinary haemangioma, EH tends to develop during adult life. The majority of angiosarcomas are fatal, whereas EH appears to have a recurrence rate of 10% and a metastatic rate of 20%. In addition, the histological features, which appear to be useful in predicting metastatic potential, include the prevalence of mitotic figures and the degree of cellular pleomorphism. One of the characteristics of this tumour is its capacity to maintain endothelial markers, including junctional attachments, pinocytotic vessels, Weibel-Palade bodies, and factor VIII-associated protein [3, 5, 7]. If possible, the treatment of choice for EH is complete resection. There is little experience with irradiation or chemotherapy.

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