

# The first reported case of epithelioid haemangioma arising from the pleura

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## Abstract

Epithelioid haemangioma is a rare, benign, vascular tumour of unknown cause, mostly found in the subcutaneous tissue of the head and neck. Here we report the first case of an epithelioid haemangioma arising from the pleura. The tumour originated from the parietal pleura in the left chest and was resected with thoroscopic assistance with the adjacent chest wall. Pathological examination revealed a mixture of lymphoid and vascular elements with inflammatory cell infiltrate typical of epithelioid haemangioma. Our case demonstrates that although rare, epithelioid haemangioma is a differential for a pleural vascular tumour.

**Keywords:** Epithelioid haemangioma; pleura, thoracic surgery, IgG4, angiolymphoid hyperplasia, eosinophilia, benign, vascular

## Introduction

Epithelioid haemangioma or angiolymphoid hyperplasia with eosinophilia is a rare, benign, vascular lesion, reported most commonly in subcutaneous tissue of the head and neck [1,2]. Its features include proliferation of capillary vessels with epithelioid endothelial cells lining them with an inflammatory cell infiltrate [1-3]. Reports exist of epithelioid haemangiomas arising from the lung, right atrium and radial artery [4-6] however, to the best of our knowledge; there are no previous reports of a pleural origin. Treatment of epithelioid haemangioma involves surgical excision. The prognosis is generally very good although recurrence occurs in one third of patients [1]. There is some debate as to whether these tumours are a benign neoplasia or a reactive process secondary to trauma [1,3]. We report a case of a pleural derived epithelioid haemangioma in a 55 year old lady.

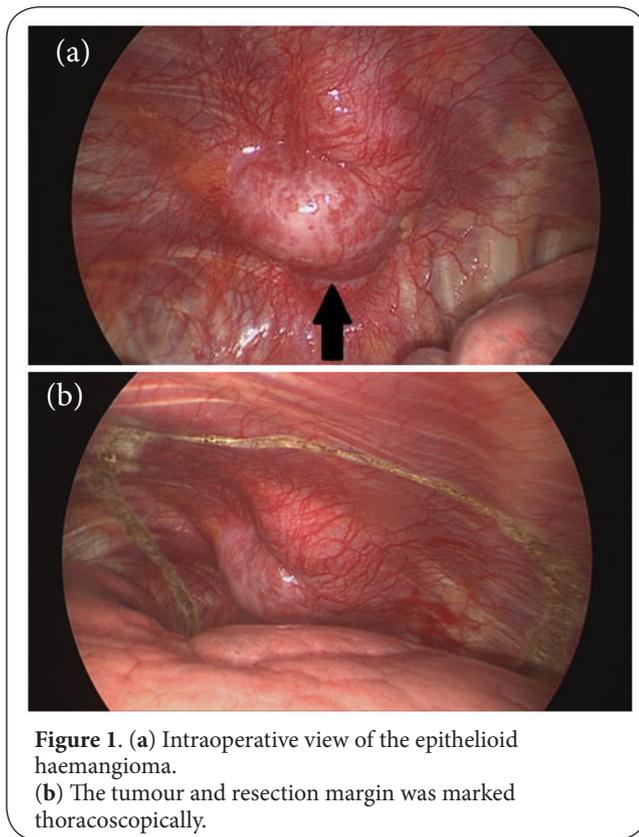
## Case presentation

A 55 year old lady with a background of emphysema, presented with a persistent cough. Chest radiograph revealed left upper zone opacity and a subsequent computed tomography scan revealed a lobulated subpleural soft tissue mass containing several prominent vessels anterior to the fourth rib posteriorly. The mass measured approximately 5x3x3 cm. Subsequent

magnetic resonance imaging (MRI) of the chest demonstrated a low T1 signal and high STIR signal of the lesion, with intense post contrast enhancement and suggestion that the lesion invaded the chest wall posteriorly. There was associated high signal STIR of the bone marrow of the 4th rib suggestive of bone infiltration. The mass extended to the level of the left T4 neural foramen, although there was no evidence of invasion or dilatation of the foramen. The appearances were reported to be in keeping with a malignant mass, such as a malignant nerve sheath tumour or sarcoma. A second MRI focused on the spine reported no involvement of the foramen, and supported the previous differential diagnosis of a malignant tumour. In contrast a positron emission tomography scan demonstrated mild to moderate uptake (SUV 3.3) consistent with a benign mass.

## Operative management

The patient was placed in the right lateral decubitus position and thoroscopic access was employed to examine the tumour (Figure 1) and delineate the resection margins (Figure 1b). The intercostal muscles were divided thoroscopically to allow for easy visualisation of the appropriate margins at external examination. A posterior-lateral thoracotomy incision was made and the involved sections of the 3<sup>rd</sup>, 4<sup>th</sup> and 5<sup>th</sup> ribs were divided as demarcated by the thoroscopic division of the intercostal

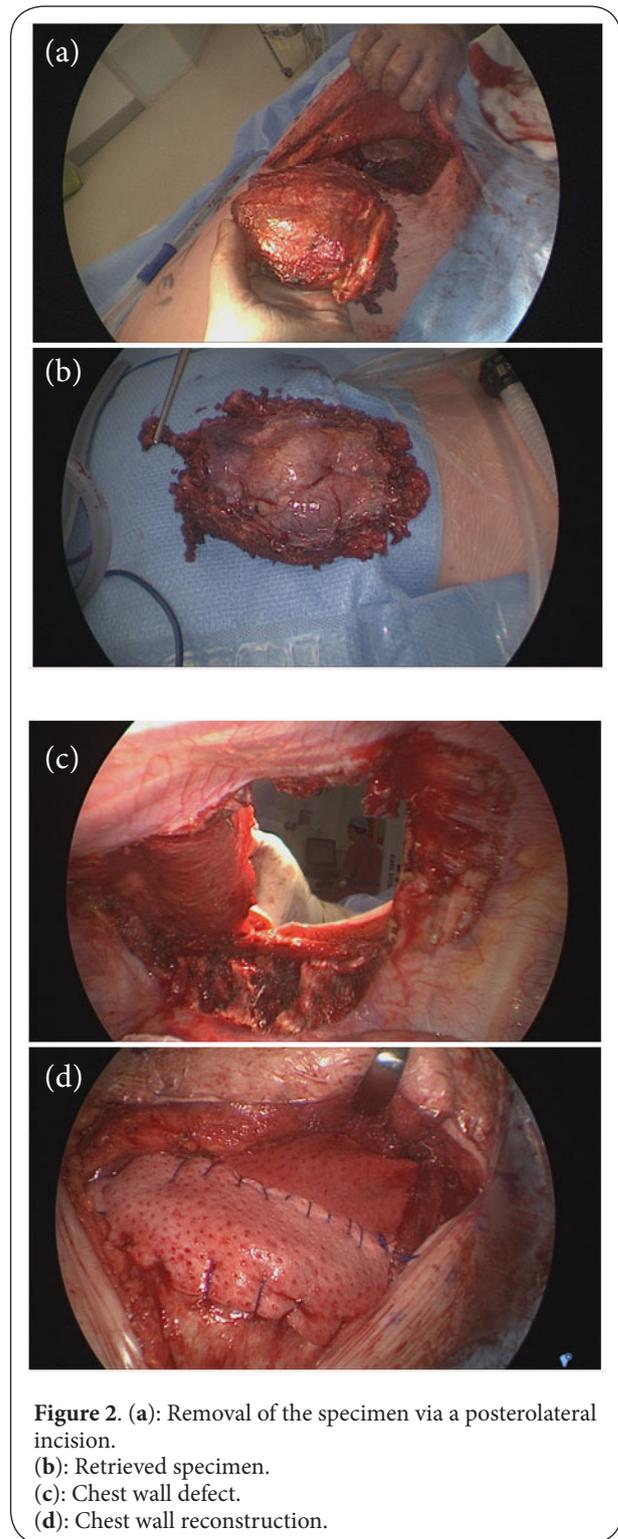


muscles. The specimen was removed en-bloc (**Figures 2a** and **2b**) and the resulting chest wall defect (**Figure 2c**) was repaired with a Permacol™ mesh (**Figure 2d**).

### Histological examination

A 35x30x15mm, well defined, irregularly shaped tumour and two irregularly shaped fibrous nodules measuring maximum of 15mm were removed (**Figure 3**). Histology slides prepared with haematoxylin and eosin stain showed a nodular lesion with features suggestive of an epithelioid haemangioma characterised by a mix of lymphoid and vascular elements. The lymphoid component comprised of lymphoid follicles with germinal centres with an inflammatory cell infiltrate rich in eosinophils, plasma cells and lymphoid cells. The vascular component consists of small to medium sized vessels, lined by epithelioid endothelial cells with abundant cytoplasm, surrounded by a mild perivascular fibrosis. IgG4 positivity was also demonstrated by immunohistochemistry using the RM120 clone after heat retrieval for 20min followed by peroxidase block. This raised the possibility of IgG4 related disease. The tumour encroached on rib 4 but did not infiltrate it. It extended into the surrounding fat and skeletal muscle.

Recovery for surgery was uneventful and the patient was discharged on post-operative day four. Serum levels of IgG4 were found to be normal which made the diagnosis of IgG4 disease very unlikely. Following discharge the patient developed



worsening breathlessness and lost 5kg in weight, following extensive investigation this was felt to be an exacerbation of her underlying chronic obstructive pulmonary disease.

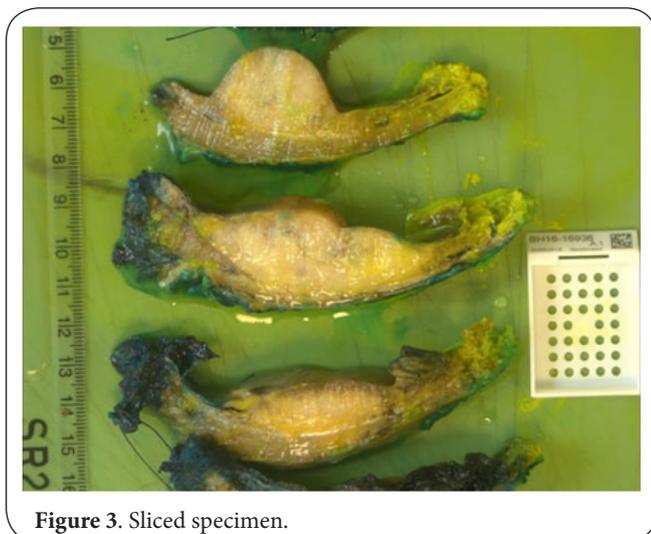


Figure 3. Sliced specimen.

## Discussion

An epithelioid haemangioma (EH) is described as a benign vascular tumour with well-formed but often immature vessels, which is lined by plump epithelioid endothelial cells with amphiphilic or eosinophilic cytoplasm and a large nucleus. Most cases have a prominent inflammatory component. The tumours affect a wide age range, peaking in the 3<sup>rd</sup> to 5<sup>th</sup> decades and have a higher incidence in women [7]. EHs are most commonly found in the subcutaneous tissue of the head and neck [1] and the distal extremities, where they generally present as purple or brown subcutaneous nodules or papules. There are a small number of reports of origin from other tissues, including the lungs, right atrium and radial artery [4-6]. However, this case - to our knowledge - is the first report of an EH arising from the pleura. The aetiology of EHs is unclear, there remains controversy as to whether the lesions arise in areas of previous trauma or are of true neoplastic origin [3]. It was originally thought that these lesions were a stage of Kimura's disease, but they are now considered to be two histologically distinctive entities [8]. These tumours can be locally invasive and rarely metastasise. This finding is a rare but important differential for a pleural tumour, as its benign nature may influence management choices for the patient.

## Competing interests

The authors declare that they have no competing interests.

## Authors' contributions

Authors' contributions	JS	UA	RD	EI
Research concept and design	✓	✓	✓	✓
Collection and/or assembly of data	✓	✓	✓	✓
Data analysis and interpretation	✓	✓	✓	✓
Writing the article	✓	✓	✓	✓
Critical revision of the article	✓	✓	✓	✓
Final approval of article	✓	✓	✓	✓
Statistical analysis	✓	✓	✓	✓

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