Case Report

Sclerosing Hemangioma of the Lung: A Benign Tumour with Potential for Malignancy?

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Pulmonary sclerosing hemangioma represents a rare neoplasm with variable potential for progression. This case report of a 35-year-old female with left-sided thoracic pain. Computed tomography revealed a centrally located, well-circumscribed and partially calcified lesion. Intraoperative findings were suggestive of a carcinoid tumour. The tumour was completely removed by lobectomy followed by systematic lymphadenectomy. The histopathological analysis revealed a sclerosing hemangioma, a rare benign neoplasm. Sclerosing hemangiomas (SHs) are true neoplasms derived from alveolar pneumocytes. However, little data is available on the potential malignant behaviour, such as lymphnode metastases, local recurrence, and the appearance of SH's. Generally, wedge resection is justified in the majority of cases, but in cases of uncertain intraoperative diagnosis, anatomic resection with systematic lymphadenectomy is recommended. (Ann Thorac Cardiovasc Surg 2006; 12: 352–4)

Key words: benign neoplasm, lung

Introduction

Sclerosing hemangioma (SH) of the lung was first described by Liebow and Hubbel.[1)] This rare pulmonary neoplasm predominantly affects females over 50 years of age. Typically, there is a solitary, generally asymptomatic, well-described lesion located in the periphery of the lung. In a series of 45 benign neoplasm, Sugio et al. found 10 tumors to be a SH, the second most common benign tumour behind hamartochondroma.[2)] While this tumour entity is more common in Asia, very little clinical data exists from Europe and the United States.

Histologically, there are epithelioid (solid), papillary, sclerotic and hemorrhagic patterns. This tumour was originally thought to be a lesion derived from vascular structures due to its rich content of blood vessels. However, immunochemical analysis showed clear evidence that SH derives from primitive respiratory epithelium. Alternatively, the term alveolar pneumocytoma is also used.

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Case Report

We describe the case of a 35-year-old female, who consulted a local hospital with cardiocirculatory instability and left-sided thoracic pain. The patient complained of a recurrent stabbing pain in the left thorax over the previous 2 years. She had an otherwise uneventful history. She worked as a mechanic with exposure to several metallic dusts over the last few years and had a smoking history of 17 pack years. The family history was positive for tuberculosis in her grandmother.

Lung embolism and myocardial infarction were ruled out by ECG, chest X-ray and laboratory parameters as well as a computed tomography of the chest.

However, the computed tomography of the chest revealed a tumour in the aortopulmonal window with little but homogenous contrast media enhancement and possible small calcifications (Fig. 1). There was no evidence of infiltration of the surrounding tissue or pathologically enlarged lymph nodes. The patient was then transferred to our hospital for further evaluation.

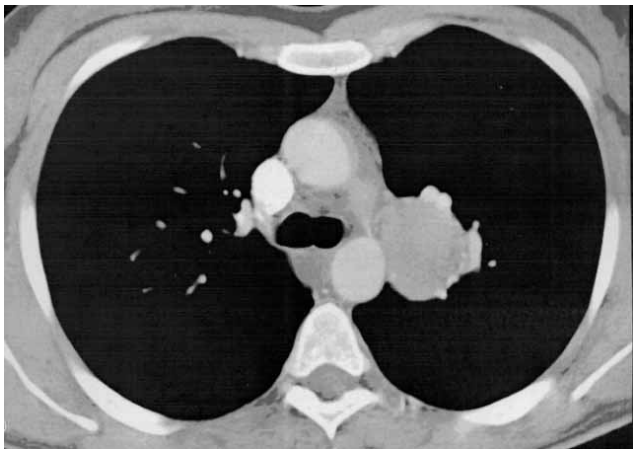

Fig. 1. Thoracic computed tomography of a 35-year-old female with a left-sided paraaortic tumour partially calcified in the periphery.

We did not take a biopsy prior to operation because, this tumour was thought to be benign and due to the higher risk of complications occurring when performing a CT-guided biopsy.

Bronchoscopy performed before the operation was normal. Exploratory thoracotomy showed an epihilar, well-perfused tumour of elastic consistency. Histology of the tumour could not be determined clearly by frozen section analysis. However, macroscopically, the lesion was suggestive for a carcinoid tumour. Therefore, a lobectomy of the left upper lobe was performed followed by a systematic lymphadenectomy.

Histological examination of the specimen showed a tumour containing cells and vessels, built from superficial cuboid epithelium and round "stroma''-cells. Immunohistology revealed the diagnosis of SH with a partially cavernous pattern with some hemorrhagic areas. The stroma cells showed a typical expression of TTF-1 and were negative for cytokeratin-markers. CD34 was not expressed by the stroma cells, but revealed a high vessel density. The proliferation index was less than 1%. Thus, the diagnosis of a sclerosing hemangioma with solid and hemorrhagic growth patterns was verified.

Comment

SH of the lung is a rare tumour without a definitive classification. However, a unique and typical feature of SHs is that they typically occur in females over 40 years of age and they are asymptomatic until the time of diagnosis.[3,4)]

SHs of the lung are mainly found incidentally (80%). Symptoms are due to enlargement of the tumour and compression of surrounding tissue. The described patient had atypical left-sided thoracic pain. However, since there was no association between the tumour and the intercostal nerves or other surrounding structures, these complaints were probably not related to the tumour.

Differential diagnosis of SH should include hamartoma, cavernous hemangioma, inflammatory lesions, arteriovenous malformations, malignant teratomas or angiosarcomas.

Chest X-ray reveals SH as a sharply marked, well defined lesion. Characteristically on the computed tomography, SHs enhance homogenous contrast media with little peripheral calcifications and there is no infiltration of the surrounding tissue.

In addition to CT, a FDG-PET is possibly able to differentiate between benign and low-grade malignant lesions and can thus help in planning an operation.[5)]

SHs of the lung are considered as neoplasms with a rich vascular structure. Due to the large amount of vascular structures, this tumour entity was thought to be of vascular origin. However, according to current understanding, it is a benign proliferation of primitive respiratory epithelium. Using antibodies against thyroid transcription factor-1 (TTF-1) and epithelial membrane antigen (EMA) type 2 pneumocytes were used to identify the origin of SH.[6,7)]

Macroscopically, SHs were found as well-circumscribed and partially cystic tumours. Histologically, this tumour is rich in vascular structures, partially hemorrhagic, papillary folded with cuboid superficial proliferating epithelium in between collagen fibers and round "stroma"-cells (Fig. 2). Atypical cells as well as mitosis are rarely seen, confirming its mostly benign behavior. In individual cases, adenocarcinoma-like cells are found within the tumour, a finding that is not detectable prior to operation, so in addition, one has to be aware of the presence of lymph node metastasis.[3,8)]

In conclusion, as soon as the indication for explorative thoracotomy becomes apparent in symptomatic patients, thoracotomy and wedge resection is the treatment of choice for SH as it allows exact diagnosis. In cases of uncertain diagnosis by intraoperative frozen section analysis and since there is only little evidence about the behaviour of metastasing and local recurrence of SH,[4,8)] anatomic resection with systematic lymph node dissection is mandatory.

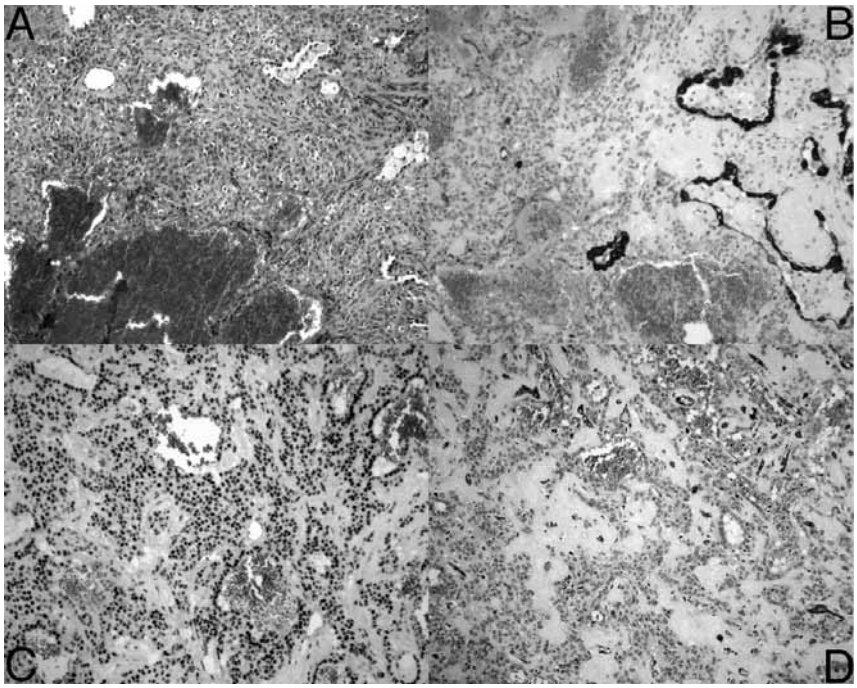

Fig. 2. Histological specimen of SH. (**A**) HE stain showing a partially hemorrhagic tumour. Immunohistochemistry revealing a negativity of the stroma cells for cytokeratin (**B**) while expression of TTF-1 (**C**). CD34 is not expressed by the solid parts of the tumour, but only stains vascular endothelium (**D**).

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