

Spindle cell sarcoma: a rare cause of a spontaneous pneumothorax

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Introduction

A secondary spontaneous pneumothorax can be associated with a variety of lung disorders such as emphysema, infections, cystic fibrosis, tuberculosis, and neoplasms. Malignancy-related secondary spontaneous pneumothoraces are rare events accounting for 0.05–1% of all pneumothoraces [1,2] and only on rare occasions this is the presenting feature. Primary tumours, usually cavitated, are less common cause of pneumothorax than secondary [1].

Primary spindle cell sarcoma is an extremely rare entity. It is a type of a connective tissue tumour, which has been reported in a variety of body parts, most commonly in the skin, muscles, and surrounding organs [3].

We present the case of an asymptomatic spontaneous pneumothorax which led to the diagnosis of a primary spindle cell sarcoma of the pleural. We review the literature and highlight the peculiarity of this case.

Case Report

A 71-year-old-woman had a mechanical fall, hurt her shoulder, and her chest radiograph showed the incidental

Abstract

Secondary spontaneous pneumothoraces are associated with a variety of lung disorders. Malignancy is a rare cause of a pneumothorax and rarely the presenting feature. Cavitating primary tumours are less frequent causes of pneumothoraces than secondary tumours. We present the case of an asymptomatic spontaneous pneumothorax in a 71-year-old never-smoking woman. Her right apical pneumothorax was diagnosed by chance on a chest radiograph for a fall. Her final diagnosis was a primary spindle cell sarcoma of the pleura. This case demonstrates the need to explore the underlying causes of a non-resolving pneumothorax. The early diagnosis of her sarcoma allowed curative treatment.

finding of a small right-sided apical pneumothorax. This pneumothorax failed to resolve over a period of nine months of conservative management.

Clinical examination nine months later showed her well with a weight of 70 kg, height of 156 cm, and body mass index (BMI) of 29. She had some hyper-resonance in the right upper chest and no other relevant abnormalities. Her past medical history includes intermittent migraines, hypertension, and non-insulin-dependent diabetes, treated with metformin 500 mg twice daily. She had never smoked.

Her chest radiograph showed a persistent pneumothorax. A computed tomography (CT) confirmed the pneumothorax and showed a small cystic-like lesion in the right upper lobe (Fig. 1). Her CT scan revealed no other features suggesting the underlying lung disease.

She underwent a right-sided video-assisted thoracoscopic surgery (VATS) with a wedge resection of the cystic abnormality in her right upper lobe and a surgical pleurodeses. This led to a complete and persistent resolution of her pneumothorax. The histology of the resected lesion revealed a synovial spindle cell sarcoma with spindle cells extending from the pleural into the lung tissues and spindle cells arranged in intersecting fascicles, with sclerotic components and epithelial

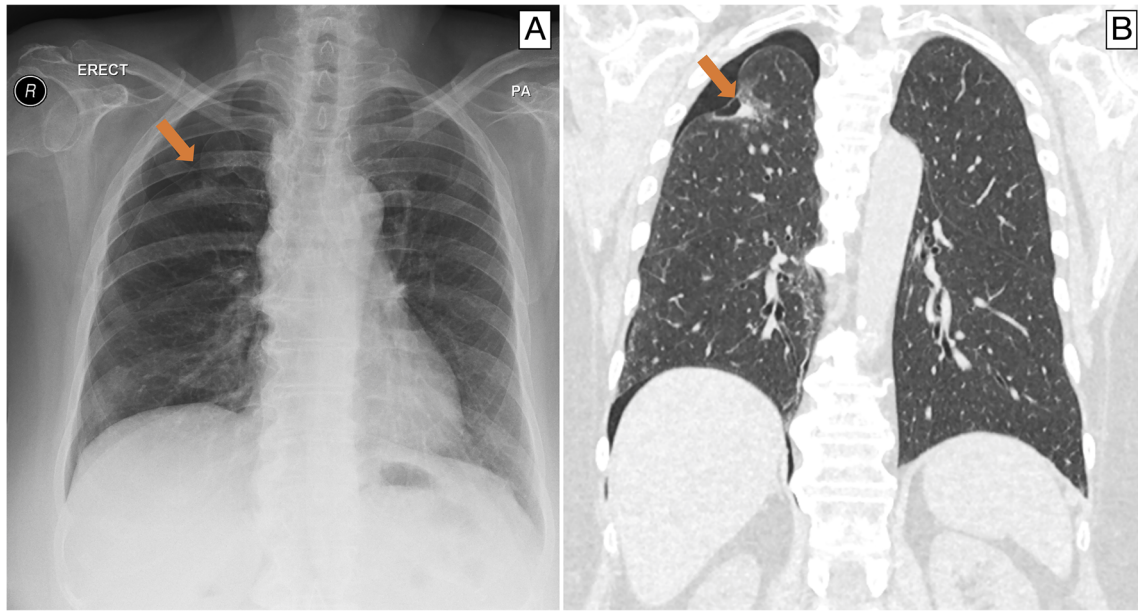


Figure 1. Chest imaging. (A) Chest radiograph showing right apical pneumothorax. (B) Computed tomography (CT) scan showing right apical pneumothorax and a cystic-like lesion in the right upper lobe.

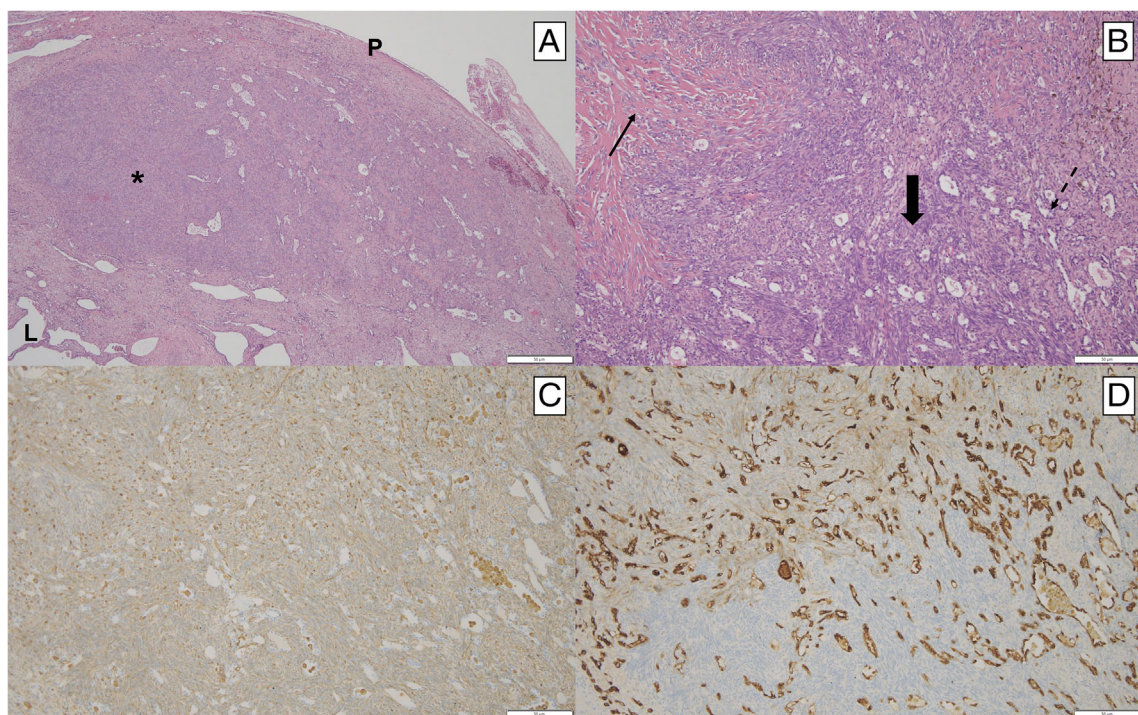


Figure 2. Synovial spindle cell sarcoma. (A) Tumour cross-section revealing tumour (*) extending from the pleura (P) into the lung tissue (L) (haematoxylin and eosin (H&E) stained, 40x). (B) Tumour composed of short spindle cells arranged in intersecting fascicles (thick arrow), sclerotic component (thin arrow), and epithelial glandular elements (interrupted arrow) (H&E stained, 100x). (C) Expression of vimentin (100x). (D) Expression of epithelial membrane antigen (EMA) (100x).

glandular elements. The immunohistochemistry was positive for vimentin and epithelial membrane antigen; AE13 and TTF1 were focally positive, and S100, SMA, desmin, and Stat6 were negative (Fig. 2). This was confirmed by the SS18 arrangement on fluorescence in situ hybridization (FISH).

After a whole-body positron emission tomography (PET) scan revealed no other sites of tumour spread, she underwent a completion right upper lobectomy about 18 months following her initial presentation.

She has remained asymptomatic and physically active for three years so far with no recurrence of her tumour or pneumothorax.

Discussion

A synovial spindle cell sarcoma is an extraordinarily rare tumour, which occurs commonly in white people in their fifth to seventh decades of life without gender predominance [4].

In a review of 153 cases of pneumothorax complicating sarcoma, only 15% were asymptomatic and had pneumothoraces discovered by screening chest radiographs. Spindle cell sarcomas were noted in 15.7% of these cancers. Typical appearance on the screening chest radiographs included multiple nodules (48.4%), cavitary or cystic lesions (25.8%), and other pleural abnormalities (11.7%). A pneumothorax complicating a sarcoma is normally associated with frequent recurrences and an increased mortality: only 8.6% of patients were alive two years after the initial diagnosis [5].

The mechanism of spontaneous pneumothorax due to spindle cell sarcoma is unknown. However, it is well known that in most patients the spontaneous pneumothorax results from the rupture of bullae or blebs. In this case, it probably results from the normal anatomic structure disruption due to the sarcoma and consequent rupture of visceral pleura. The patient was asymptomatic and the pneumothorax was discussed as an incidental finding, which added to the delay between diagnosis and definite treatment.

This case combines two exceedingly rare scenarios into a positive patient outcome. First, the patient had a primary pleural spindle cell sarcoma, whereas most published cases seem to describe secondary, metastatic tumour involvements.

Second, the tumour was diagnosed following an incidentally discovered, asymptomatic pneumothorax. The combination of these two events led to the atypical long survival compared to the previously published cases.

Our case underlines the value in exploring the possible underlying causes of spontaneous pneumothoraces. The early diagnosis of the primary sarcoma allowed an effective curative treatment with a good prognosis.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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Author Contribution Statement

Ana Pedroso: Conception, acquisition of information, analysis or interpretation of data, drafting the manuscript. Lutz Beckert: Review of the work and final approval of the version to be published.

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