Clinical Case Report

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Extrapleural solitary fibrous tumor in thymic area
A case report

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Abstract

Rationale: Solitary fibrous tumor (SFT) is a rare benign soft tissue mesenchymal neoplasm. There have been a few reports of extrapleural SFTs although it can occur anywhere in the body.

Patient concerns: A 30-year-old male presented with an anterior neck mass since one month.

Diagnoses: Based on physical and radiologic examination, preliminary differential diagnosis was thymic neoplasm or intrathoracic goiter.

Interventions: We performed surgical excision of the neck mass.

Outcomes: The histopathological examination revealed an extrapleural SFT

Lessons: We present a case of extrapleural SFT in the thymus which was misdiagnosed as thymoma based on radiologic findings.

Abbreviations: CT = computed tomography, SFT = solitary fibrous tumor.

Keywords: extrapleural, solitary fibrous tumors, thymus gland

Key Points

- Extrapleural solitary fibrous tumor (SFT), first described in 1931, is a rare and mostly benign spindle cell tumor.
- The radiologic characteristic of extrapleural SFT is a well-defined enhancing mass, and immunohistochemical staining showed positivity for CD34 and STAT6.
- We present extremely rare case of extrapleural SFT in the thymic area that had a benign course and was treated definitively with surgical excision.

1. Introduction

Solitary fibrous tumor (SFT) is a benign soft tissue mesenchymal neoplasm, which was first described in 1931 by Klemperer and Rabin.[1] It is a rare tumor and is typically found within the pleura. There have been a few reports of extrapleural SFTs.[2] Herein, we present a case of extrapleural SFT in the thymus which was misdiagnosed as thymoma based on radiologic findings.

2. Case report

A 30-year-old male presented with an anterior neck mass since 1 month. The patient had no significant medical history except that he had recently lost a lot of weight through vigorous dieting. The patient had an anterior soft huge mass without tenderness. Physical examination revealed no enlarged lymph nodes. Computed tomography (CT) scan of the neck revealed a 6.5 cm, well-enhanced soft tissue tumor with internal necrotic change in the left anterior lower neck, just inferior to the thyroid

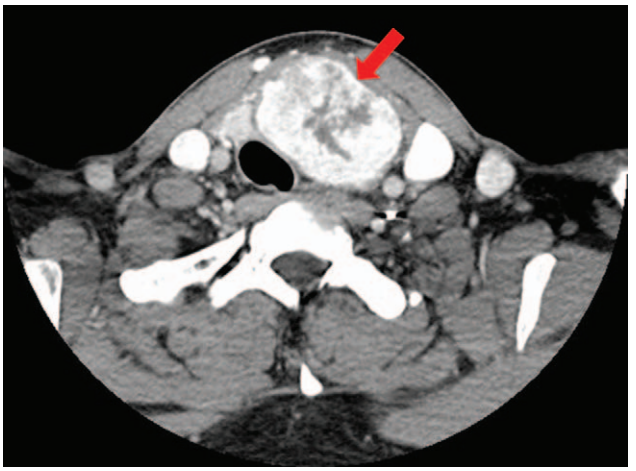

Figure 1. Axial computed tomography scan of the neck shows a 6.5-cm sized well-enhanced soft tissue tumor with internal necrotic change in the left anterior lower neck, just inferior to the thyroid.

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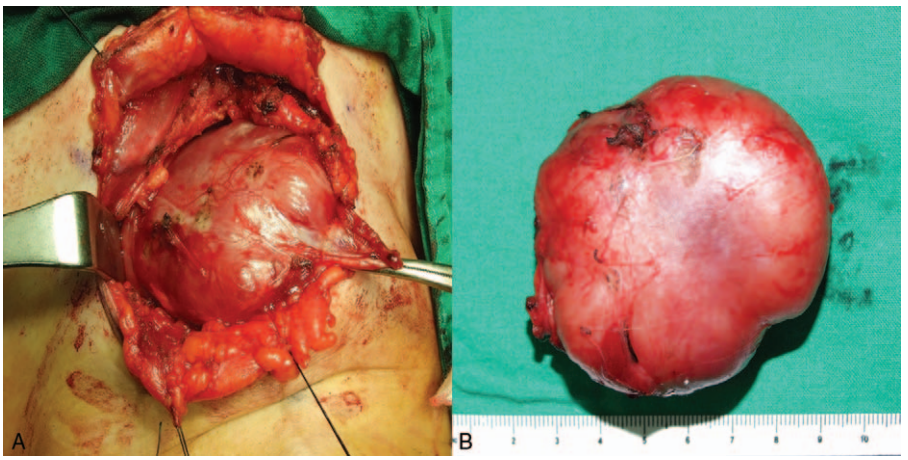

Figure 2. Intraoperative finding shows a 6.5-cm sized huge mass, which was carefully separated from the surrounding structures (A). Surgical specimen (B).

and a few enlarged lymph nodes in both carotid spaces (Fig. 1). Fine-needle aspiration cytology was performed under ultrasound guidance; however, an adequate cell harvest was not obtained except benign inflammatory cells.

Based on these observations, preliminary differential diagnosis was thymic neoplasm or intrathoracic goiter. Two months after diagnosed lesion, we performed excision of the neck mass. During surgery, a transverse incision was made over the anterior neck. The dissection proceeded through the thyroid level. A mass was pushing the thyroid toward the trachea from the left side. A 6.5-cm sized huge mass was carefully separated from the surrounding structures, and it was totally resected (Fig. 2). When we cut the mass, a well-encapsulated solid tumor with focal necrosis was observed. Pathological examination of the lesion revealed extrapleural SFT, which showed CD34 positivity and STAT6 positivity (Fig. 3). The postoperative course was

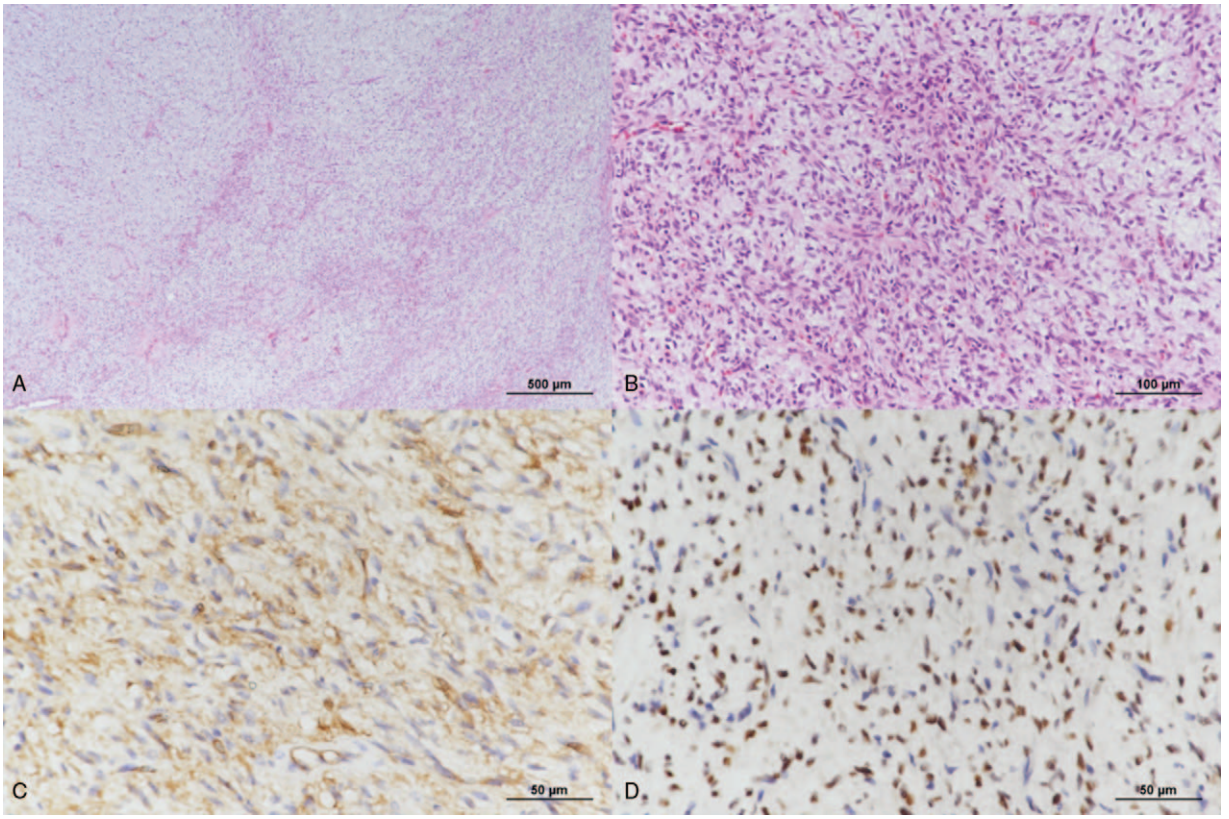

Figure 3. Pathologic findings of the resected tumor. (A) The low-power view of the tumor shows a mixture of hypocellular and hypercellular areas intervened by arborizing, hyalinized vessels (hematoxylin and eosin, original magnification ×40). (B) Tumor cells are predominantly spindle-shaped with pale cytoplasm and separated by thin-walled vessels (hematoxylin and eosin, original magnification ×200). (C) Immunohistochemistry using CD34 antibody shows diffuse cytoplasmic positivity (immunohistochemistry, original magnification ×400). (D) STAT6 immunohistochemistry, the confirmative marker for diagnosis of solitary fibrous tumor, displays strong nuclear reactivity in tumor cells (immunohistochemistry, original magnification ×400).

uneventful. At the last follow-up, there has been no recurrence, and the patient remains asymptomatic. This study was approved by the institutional review board of the Chonnam National University Hwasun Hospital. Informed consent was given by the patient.

3. Discussion

Extrapleural SFT, first described in 1931, is a rare and mostly benign spindle cell tumor.[3] It can occur at various sites, resulting in various symptoms. Usually SFTs are slow-growing tumors with favorable prognosis, although there have been a small number of malignant cases.[4] Although its lineage is still uncertain, a recent study has suggested a myofibroblastic/fibroblastic lineage due to its strong CD34 immunoreactivity, patchy actin positivity, and cytoplasmic dense bodies.[5] Radiologic characteristics of SFT have been described as a well-defined enhancing mass on both CT and magnetic resonance imaging.[6] A previous study assessing the gross section of the tumor showed well-circumscribed, lobulated, rubbery masses with a tan-pink surface and whitish-gray cut surface, which was consistent with our case.[7]

SFTs can cause various symptoms such as diplopia, nasal obstruction, or even difficulty in breathing when they occur in the thoracic cavity.[8] However, commonly SFTs present as slow-growing painless masses when they occur in the head and neck.[6] Our patient had an anterior neck mass since 1 month; however, the mass may have been detected recently due to the recent weight loss although it had been present for some time.

Although there are proposed diagnostic criteria for SFT, there are no definitive criteria.[9] SFT can occur in diverse circumscribed sites with scanty cytoplasm admixed with collagen.[9] CD34 stains various spindle cell neoplasms, and it has been helpful but is not specific in making a differential diagnosis.[4] In a recent study, the NAB2/STAT6 fusion gene has been identified in RNA and DNA from SFTs.[8] Along with radiologic characteristics, a well-defined enhancing mass, and gross features of the mass, immunohistochemical staining showed positivity for CD34 and STAT6, which is consistent with previous studies.[8]

The criteria of malignancy for SFT were not established. Clinicians have evaluated the histological parameters, including high cellularity, frequent mitotic activity, nuclear pleomorphism, and presence of necrosis; however, they have failed to prove the correlation for unfavorable prognosis for SFT.[4] In addition, Yokoi et al[10] suggested that the expression of p53 protein might be related to malignant clinical behavior and histologic features.

Even though SFTs show a benign histology and behavior, it does not always correlate with clinical outcomes.[3] Therefore, complete surgical excision is the treatment of choice.[3] In this case, complete excision was performed while preserving the surrounding anatomy. Surgical respectability is the single most important determinant.[3] On the other hand, in some reports, even when complete surgical excision was performed, a number of patients had local relapse or distant spread.[3] Therefore, regular patient follow-up is mandatory. We did not experience any complications related to the surgery or relapse.

In conclusion, we present extremely rare case of extrapleural SFT in the thymic area that had a benign course and was treated definitively with surgical excision.

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