Case Report

A Rare Case of Ancient Schwannoma in the Anterior Mediastinum: a Case Report

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Abstract

Ancient schwannoma is a rare variant of schwannoma that presents histological features mimicking malignancy, often leading to diagnostic challenges. This benign neurogenic tumor originates from Schwann cells of the peripheral nervous system and can develop throughout the sympathetic and parasympathetic nervous systems. Neurofibromas and schwannomas are among the most common benign neurogenic tumors, but their occurrence in the anterior chest wall, particularly in the mediastinum, is exceedingly rare, with limited cases documented in the literature. We report the case of a 45-year-old female who presented with a 6-month history of progressive shortness of breath and chest pain. The final histopathological examination and IHC confirms the lesion to be an ancient schwannoma. These tumors, characterized by distinctive degenerative changes, often present in atypical locations and are associated with diverse clinical manifestations and unpredictable behavior. Thoracoscopic excision is preferred for small to moderate mediastinal schwannomas due to its minimally invasive nature. However, for large tumors, thoracotomy is often required to ensure complete and safe resection. As these tumors are benign, radiotherapy or chemotherapy is usually not needed. Diagnosis relies on histopathology, though imaging aids in preoperative evaluation. No recurrence was observed in this case.

Keywords: Schwannoma; Benign tumor; Neural tumor; Sternotomy; Mediastinum

Introduction

The mediastinum, a complex anatomical compartment, is susceptible to a broad spectrum of pathological conditions, including both non-neoplastic and neoplastic processes. Mediastinal neoplasms, which may be benign, malignant, or metastatic, are categorized as primary when they arise from structures within the mediastinum or from tissues that traverse this area during embryonic development [1]. Neurogenic tumors, such as neurofibromas and schwannomas, are benign nerve sheath tumors that can develop anywhere in the body along peripheral nerves [2]. While these tumors are often linked to neurofibromatosis type 1 or 2 [3]. They can occasionally present as solitary lesions on the chest wall or involve the pleura or mediastinum [4, 5], with schwannomas being notably rare in the anterior mediastinum [6]. Schwannomas, although typically benign, pose significant clinical challenges due to their potential to cause morbidity through compression of adjacent structures [7]. These tumors are most frequently observed in the head, neck, and posterior mediastinum, but cases involving the anterior mediastinum, especially those arising from intercostal nerves, are extremely uncommon [8]. Imaging modalities like computed tomography (CT) and magnetic resonance imaging (MRI) play a crucial role in assessing these tumors [9], but definitive diagnosis depends on histopathological examination, which reveals characteristic features such as Verocay bodies and Antoni A and B regions. In this case report, we describe a 45-year-old female who presented with a 6-month history of shortness of breath and chest pain. Imaging studies revealed a mass in the anterior mediastinum, which was later confirmed through histopathology to be an ancient schwannoma. This case highlights the importance of considering rare neurogenic tumors in the differential diagnosis of mediastinal masses and the challenges they pose in clinical management.

Case Report

A 45-year-old female presented with a 6-month history of progressive shortness of breath and chest pain but no associated cough, hemoptysis, fever, weight loss, or other systemic symptoms. Her medical history was unremarkable, with no history of hypertension, diabetes mellitus, chronic heart disease, or asthma. She was not on any long-term medications, and there was no relevant family history. On physical examination, her vital signs were stable, and auscultation revealed decreased breath sounds in the anterior right thoracic region, though no wheezes or crackles were noted. A chest X-ray showed a well-defined soft tissue mass in the anterior mediastinum, and further evaluation with contrast-enhanced CT (CECT) of the chest revealed a large, heterogeneous mass measuring approximately 10×9 cm in the anterior mediastinum with both cystic and solid components. The mass did not invade adjacent structures but compressed surrounding tissues. There was no lymphadenopathy or other abnormalities noted on imaging, and her laboratory investigations, including complete blood count, liver and renal function tests, were within normal limits. Due to the suspicion of a mediastinal tumor, a CT-guided fine-needle aspiration biopsy (FNAB) was performed but it was inconclusive. This was attributed to the limited sample tissue received and the presence of extensive degenerative changes and cytological atypia, which hinders definitive diagnosis in ancient schwannomas. Consequently, the patient underwent a median sternotomy for complete excision of the mass. Intraoperatively, a well-encapsulated, lobulated mass was identified in the anterior mediastinum, which was excised en bloc along with two adjacent lymph nodes. The excised specimen consisted of a single whitish-brown, soft to firm, globular tissue measuring 10×8.8×4.2 cm with a dull and congested outer surface. On cut sectioning, the tumor showed solid cystic areas with yellowish fluid coming out of it, along with gelatinous yellowish regions (Figure 1). Microscopic examination revealed a mixture of hypocellular and hypercellular areas composed of numerous elongated, narrow cells with tapered ends, dense chromatin, and ill-defined cytoplasm. There was evidence of nuclear palisading around fibrillary processes, and Verocay bodies were observed in the cellular areas. Other findings included cystic degeneration, hemorrhage, thick-walled hyalinized blood vessels, and chronic inflammation (Figure 2). Immunohistochemical staining demonstrated diffuse strong nuclear and cytoplasmic positivity for S100 protein, confirming the diagnosis of schwannoma (Figure 3), while the Ki-67 labeling index was approximately 5%, indicating low proliferative activity. The two lymph nodes submitted also showed features of reactive lymphoid hyperplasia without any evidence of malignancy. The patient had an uneventful recovery postoperatively and was discharged on the fifth day after surgery. She reported significant improvement in symptoms and showed no signs of recurrence during follow-up visits at 1, 3, and 6 months. This case underscores the importance of correlating clinical, radiological, histopathological, and immunohistochemical findings for an accurate diagnosis and management of mediastinal schwannomas, which, although rare, can be successfully treated with complete surgical excision.

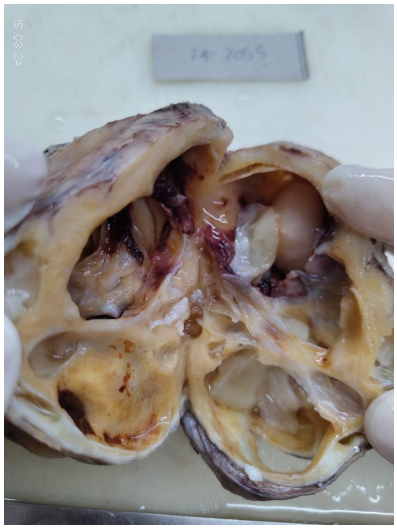

Figure 1: Grossly the tumor showed whitish-brown, soft to firm, globular mass measuring 10×8.8×4.2 cm with solid cystic areas along with gelatinous yellowish regions.

Discussion

Schwannoma, or neurilemmoma, is a typically solitary benign tumor that originates from the peripheral nerve sheath [10]. These tumors represent around 95% of all benign neurogenic mediastinal tumors derived from Schwann cells [11, 12]. Ancient schwannomas represent 0.8% of soft tissue tumors [13]. These tumors typically develop from spinal nerve roots, however they can also develop from intrathoracic nerves. Furthermore, schwannomas can be discovered in the lungs in about 0.2% of cases [14]. The clinical presentation of benign mediastinal schwannomas often includes symptoms such as cough and chest discomfort, with some patients also experiencing weight loss and hoarseness. Degenerative changes such as hemorrhage, hyalinization, cyst formation, and calcification, as seen in our case, are typical features [15]. Symptoms often

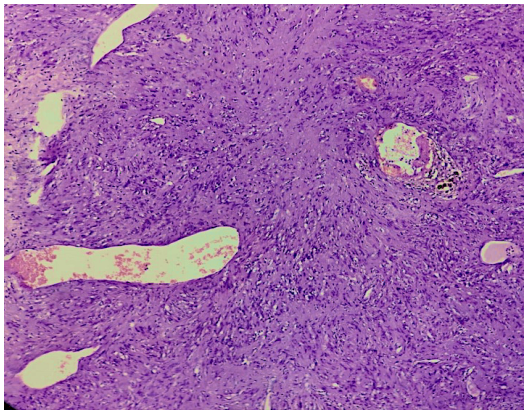

Figure 2: Mixture of hypocellular and hypercellular areas composed of numerous elongated, narrow cells with tapered ends, dense chromatin, and ill-defined cytoplasm. There was evidence of nuclear palisading around fibrillary processes. Other findings included cystic degeneration, hemorrhage, thick-walled hyalinized blood vessels, and chronic inflammation. (H&E stain, 10×)

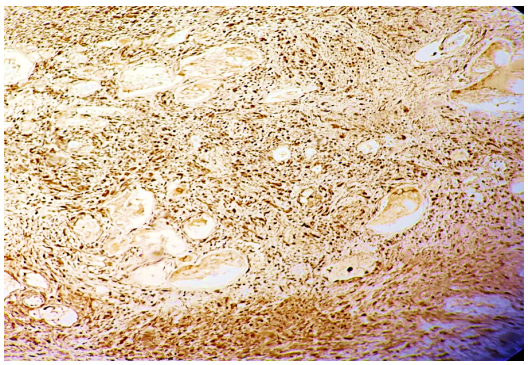

Figure 3: IHC: S100 showing nuclear and cytoplasmic positivity.

arise from the gradual enlargement of the mass, with pressure effects on nearby organs or sensory changes along the affected nerve, which was the only symptom in our patient, who maintained a good performance status. Radiological findings of soft tissue calcification and cystic areas are characteristic of ancient schwannomas and are uncommon in other schwannoma types. Radiologically, mediastinal schwannomas typically appear as well-defined lesions, occasionally exhibiting calcifications [14]. On non-contrast CT scans, they present as smooth, spherical, hypodense lesions located within the chest wall muscles. After contrast administration, these tumors can display various enhancement patterns, ranging from homogeneous to those with multiple hypodense or cystic areas. In our case, CT scan showed a sizable heterogeneous mass measuring approximately 10×9 cm located in the anterior mediastinum, comprising both cystic and solid components. Bronchoscopy can be a valuable tool in differentiating mediastinal schwannomas from primary lung cancers [15]. Histopathological examination of schwannoma shows a well-defined, encapsulated, grey-white tumor linked to nerves, often with cystic degeneration [15]. In our case, histopathology revealed a well-encapsulated tumor showing both hypercellular (Antoni A) and hypocellular (Antoni B) areas, consistent with classical schwannoma morphology described in the literature [11]. Surgical resection is the main treatment for benign neurogenic tumors, except neuroblastoma. Thoracotomy is used for larger tumors, while video-assisted thoracoscopy is ideal for smaller ones, reducing morbidity and recovery time [8]. Complete excision is usually sufficient, with no reported recurrence and excellent long-term outcomes [10].

Conclusion

In conclusion, mediastinal schwannomas, although often asymptomatic, can present with symptoms like breathlessness and chest pain, as seen in this 45-year-old female. Diagnosis relies on radiological imaging and confirmation through histopathological analysis. Surgical resection via thoracotomy is effective, especially for larger tumors, with an excellent prognosis and no reported recurrences. Distinguishing features on imaging aid in differentiating intrathoracic schwannomas from other nerve sheath tumors.

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