Ancient Schwannoma of the Anterior Chest Wall: Report of A Rare Case

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Abstract

Benign neurogenic tumors arise from cells of the peripheral nervous system. They can be found anywhere throughout the sympathetic and parasympathetic nervous system. Neurofibromas and schwannomas are two of the most common types of benign neurogenic tumors arising from nerve sheath cells. Development of these tumors on the anterior chest wall is exceedingly rare, with limited cases reported in the literature. Here is a presentation of a 44-year-old male with a one-year history of a painless lump to the anterior chest wall that turned out to be an ancient schwannoma in the final histopathology report.

Keywords: Chest wall tumor, neurogenic tumor, schwannoma, ancient schwannoma

Introduction

Neurofibromas and schwannomas are benign neurogenic nerve sheath tumors arising anywhere in the body from minor or major peripheral nerves at any level.[1] They are commonly associated with neurofibromatosis type 1 or 2.[2] In exceptional cases, neurofibromas can present as a solitary mass on the chest wall with no underlying cause**;** in other cases, they can involve the pleura or the mediastinum.[3,4] Worldwide, it affects both genders equally.[5] Age of onset is variable; however, localized lesions are commonly seen in adults between 20-40 years old. The odds of malignant transformation are extremely low.[5]

Moreover, a chest wall intercostal nerve schwannoma is rare.[6] They are seen in both genders equally between 30-60 years old.[7] To our knowledge, limited cases of chest wall neurofibroma and schwannoma arising from intercostal nerve fiber were reported.

Case Report

A healthy 44-year-old male presented with a one-year history of progressive enlargement of a painless swelling on the chest wall above the right nipple. There were no associated symptoms nor B symptoms - a group of systemic symptoms including fever, night sweats, and unintentional weight loss - and no personal or family history of malignancy.

During chest examination, a 5x5 cm, round, hard swelling extending from the right third intercostal space to the fourth intercostal space above the nipple, 3 cm lateral to the sternum, fixed to the underlying rib. There were no palpable or painful lymph nodes in the region.

Imaging with chest x-ray reported an oval shape, retrosternal lesion with no signs of sternal lesion or bony erosion. A chest CT scan revealed a 51x39.5x40.5 mm dumbbell-shaped mass lesion(HU 25-35) seen beneath the right pectoralis major muscle extending into the right hemithorax through adjacent 3rd intercostal space to lie extra-pleurally. No calcification was noted, with no enhancement after administering contrast [Figure 1].

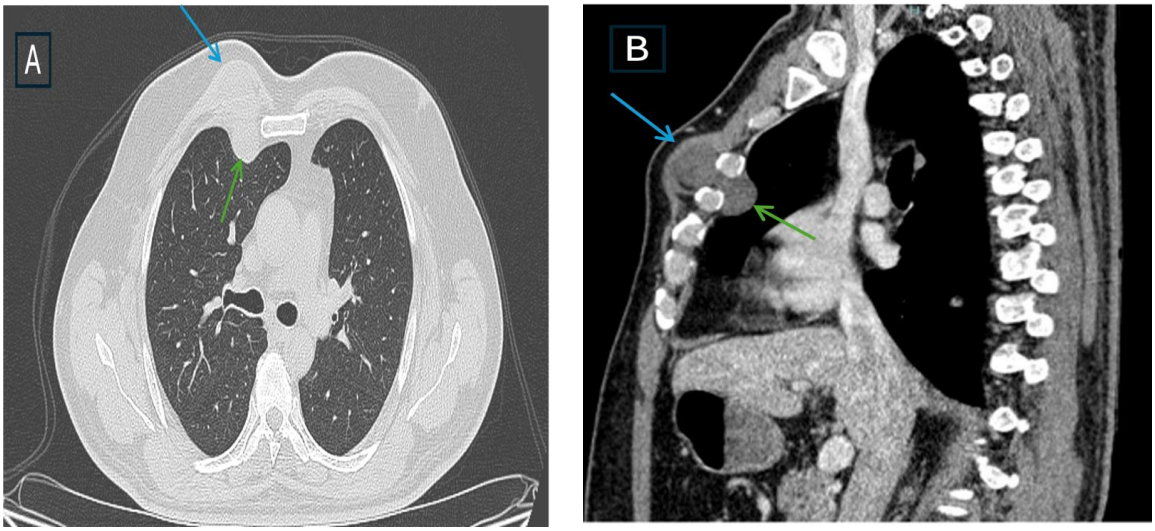

Figure 1: Cross and Sagittal sections on a CT scan showed a well-defined lesion in the anterior chest wall with extrathoracic (blue arrow) and intrathoracic (green arrow) extension between 3rd and 4th ICS.

An ultrasound-guided biopsy was done, and the result was suggestive of neurofibroma. Based on the imaging and histopathological findings, the decision was made to excise the lesion. Intra operatively, the mass was identified below the pectoral muscle [Figure 2]. The mass was then carefully dissected from surrounding structures, including the ribs and intercostal muscle. However, there was difficulty in extracting the intrathoracic part of the mass, so the decision was made to cut around 1 cm of the 3rd rib for better exposure. Two samples were sent for pathological investigation [Figure 3].

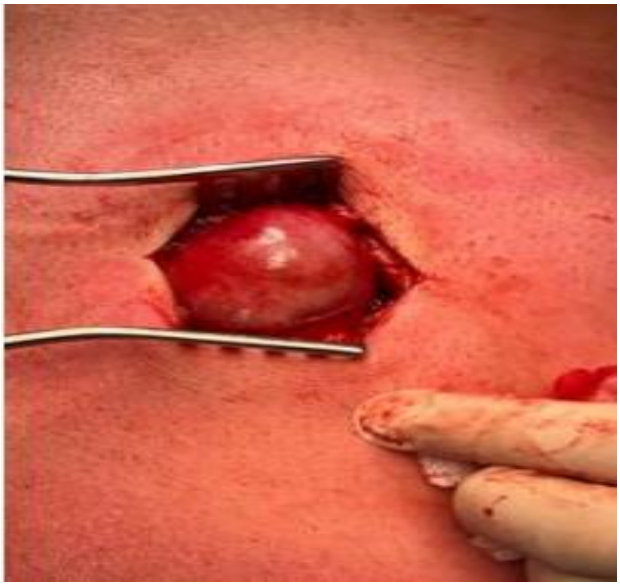

Figure 2: Intraoperative finding of a round mass below the pectoralis major muscle.

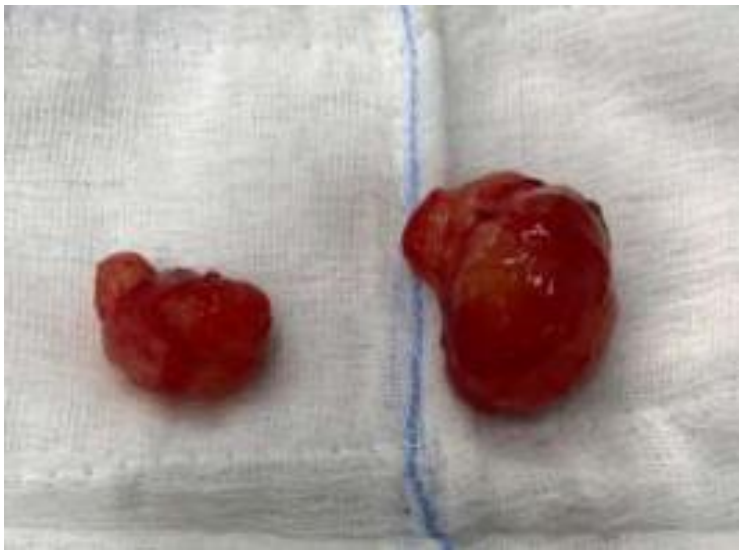

Figure 3: Specimen of 2 pieces, the larger was extra thoracic and the smaller was intrathoracic.

Postoperatively, the patient was doing fine without complications and was discharged home a few days later. During follow-up, the patient remained clinically well.

The final histopathology was reported as Ancient Schwannoma, with a microscopic finding of a circumscribed lesion with variable cellularity surrounded by a thick fibrous capsule. No malignancy was observed. Immunohistochemistry results were positive for SOX10, S100, and collagen IV. Stains for CD34, EMA, SMA, Desmin, and Caldesmon were negative.

Discussion

Tumors of the chest wall are mostly metastatic in nature, with primary chest wall lesions having an incidence of less than two percent of the population,[7,8] 45% of which originates from soft tissue, and the rest from cartilage or bone.[9] The chest wall is a rare site for benign nerve sheath tumors, with higher occurrence in the posterior mediastinum or posterior chest wall, detected as an incidental finding during radiological investigation for other complaints.[10-12] The limited literature on benign neurogenic tumors of the anterior chest describes it as a painless, palpable lump unless the tumor is large enough to result in pain due to local mass effect or neurological symptoms from dysfunction of the nerves they arise from.[12] This was seen in a case reported in India by Kale et.al of a 51-year-old lady with symptoms of chest pain and cough, attributed by pleural effusion reactionary to the presence of a pleural tumor.[10] Schwannomas are grossly solitary, firm, dumbbell-shaped, well-encapsulated, grayish lesions.[9] Less than 10% of primary tumors of the chest wall are attributed to schwannomas.[11]

Classically, a schwannoma (less commonly known as neurinoma or neurilemmoma) is a benign, slow-growing, non-recurring neoplasm occurring more commonly in individuals aged 30-60 with no gender predisposition.[11,12] As seen in 90% of cases, its occurrence is solitary and sporadic in nature. It favors the head, neck, and flexor surfaces of the upper and lower extremities, particularly the upper limbs.[10,12]

Imaging plays a vital diagnostic role. Computerized tomography (CT) and MRI are the gold-standard radiological investigations.[8,11] Typically, a schwannoma is visualized as a well-circumscribed, extrapulmonary mass with a density similar to a muscle, displacing adjacent structures without direct invasion or calcification, similar to the finding in the presented case. Larger schwannomas are more likely to show heterogeneity due to cystic degeneration or hemorrhage, as seen in 5% of cases.[13] However, a chest X-ray is often the initial investigation of choice. In our patient, the chest x-ray showed a retrosternal lesion as described initially. There was also pleural effusion in a few reported cases, including the case reported by Kale et.al.[10]

In most cases, radiography alone is insufficient for final diagnosis, so a histological evaluation is recommended. Pre-operative ultrasound-guided biopsy and immunohistochemistry were performed in this case, with initial findings suggesting neurofibroma, which was confirmed by final histopathology as Ancient Schwannoma. Pathologically, schwannomas are encapsulated benign tumors originating from the Schwann cell sheath of any nerve, with various subtypes depending on the microscopic findings.[13] It is characterized by spindle nuclei with two distinct patterns of growth: Atony A areas (hypercellular areas with nuclei palisading around a central mass of cytoplasm called Verocay bodies) and Atony B areas (hypocellular areas loosely arranged with no distinct pattern of growth). A schwannoma is termed Ancient when there is degenerative changes and diffuse hypocellular areas i.e. Consisting mostly of Atony B areas.[10]

Final immunohistochemistry results were positive for SOX10, S100, and collagen IV. However, the CD34, EMA, SMA, Desmin, and Caldesmon Stains were negative.

SOX10 transcription factor plays a role in the development and survival of the peripheral nervous system; it is highly expressed in melanocytes Schwann cells, making it a prime marker for Schwannomas. In a study evaluating 591 neurogenic and related tumors/mimics, 101 of which were represented by schwannomas, 100 out of 101 expressed SOX10, making it a highly sensitive marker.[12] Schwannomas also typically show strong expression of collagen IV and S100, which were also positive in this case.[14]

Immunohistochemistry allows differentiation between benign and malignant neurogenic tumors, most of which are negative for S100 and SOX10.[12] However, since neurofibromas are composed of Schwann cells, albeit a much smaller proportion, they also express SOX10, S100, and collagen IV, making differentiation between the two tumors difficult. However, unlike Schwannomas, neurofibroma is positive for EMA and lacks a capsule, a feature almost universal in Schwannomas; this was also consistent in our case, where the EMA was negative.[14]

Treatment of choice is predominantly with surgical resection of the tumor, with a low rate of recurrence and malignant transformation.[12] Surgical approaches include video-assisted thoracic surgery (VATS) and thoracotomy. The former is associated with a lower morbidity rate, reduced hospital stay, and fewer post-operative complications when compared to thoracotomy.[8] In cases where the tumor is large, including a case reported by Husodo S and Wati FF of a 61-year-old female with mediastinal schwannoma, resection by thoracotomy is indicated.

Schwannoma at the chest wall is a rare entity with variable imaging and biopsy findings. At present, complete surgical excision is considered diagnostic and the treatment of choice, and to date, no neoadjuvant or adjuvant therapy has been reported. A large retrospective study of 25 patients with intrathoracic neurogenic tumors was done by Natale et.al in the Thoracic Surgery Unit of the University of Campania Luigi Vanvitelli. They all underwent complete surgical excision, 72% of which were performed using VATS, and none of whom had tumor recurrence until 6 months of follow-up.[15]

Most literature describes the post-operative period as unremarkable.[3,8,13,16] There is a general consensus that benign schwannomas have an extremely low recurrence rate and malignant degeneration within 20 months following initial excision without requiring subsequent surgery, as seen in the aforementioned cases.[5,8,13,16] Nonetheless, long-term follow-up is advisable in the case of recurrence, albeit the low risk, as local recurrence can occur in the event of incomplete lesion excision.[16]

Conclusion

Solitary chest wall Schwannomas are extremely rare. The mainstay of the diagnosis is histopathology; detailed imaging is crucial and significant for management. This case elicits the importance of proper clinical assessment to prevent misdiagnosing rare cases and improve our understanding and diagnostic awareness of chest wall neurofibroma in clinical practice.

Disclosure

The authors declared no conflicts of interest. The consent from the patient was taken before Writing up the case.

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