

Case Report

Ancient Schwannoma of Chest Wall - A Rare Finding

Atul Jain¹, Ajay Suryakant Wani², Reeta Dhar³, Shilpi Sahu⁴, Puja Iyengar Ambrish²¹Assistant Professor, ²Resident, ³Professor & HOD, ⁴Associate Professor,
Department of Pathology, M.G.M Medical College & Hospital, Kamothe, Navi Mumbai.

Corresponding Author: Ajay Suryakant Wani

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ABSTRACT

Schwannoma is a slow growing tumor arising from the outer sheath of a peripheral nerve. The tumor is typically eccentric to the nerve fibres, Chest wall schwannoma accounts for 5% of all thoracic neoplasms. Ancient schwannoma constitutes 0.8 % of soft tissue tumors. The flexor surface of extremities, mediastinum, retroperitoneum, head and neck are the most common sites. Long standing and large lesions, known as ancient schwannoma, may have cystic changes, calcification, haemorrhage and fibrosis. We report a case of a chest wall ancient schwannoma in a 62year male, who initially presented with an exophytic, pedunculated growth below the left nipple. Clinically and radiologically it can be misdiagnosed as a fibrous tumor. Histopathological examination is essential for confirmation of diagnosis assisted with immunohistochemistry in few cases.

Keywords: Chest wall swelling, Verocay bodies, S-100 protein, Ancient schwannoma.

INTRODUCTION

Schwannoma is an encapsulated benign nerve sheath tumor. Ancient schwannomas are those displaying marked atypia of degenerative type. ⁽¹⁾ Schwannomas occur at all ages but most commonly occur between 20 to 50 years. ⁽¹⁾ They are usually large tumors of long duration, and a significant number are located in deep structures such as retroperitoneum and mediastinum. ⁽¹⁾ Chest wall schwannomas present as asymptomatic solitary masses. About 90% are sporadic in nature. It is a slowly growing tumor that is usually present several years before diagnosis. ⁽¹⁾ MRI & CT scan of the thorax help in better characterization of the lesion. Treatment is complete surgical excision.

CASE REPORT

A 62 year old male came with complaints of swelling on the left side of the chest since twenty five years with history of smoking since twenty years. Swelling was gradually progressive and was associated with dull aching pain. There was no history of cough, breathlessness, fever or any rash. On examination there was a solitary, freely mobile, well circumscribed swelling, soft to firm in consistency, measuring 9cm x 5cm x 3cm. No cervical or axillary lymph nodes were palpable. Systemic examination was unremarkable. Hematological and biochemical parameters were within normal limits. No MRI/CT scans were done. Patient underwent surgical resection and the swelling was sent for histopathological examination.

Gross Findings: External surface (Figure - 1, A) was covered with skin and the swelling measured 9cm x 5cm x 3 cm with an ulcerated surface showing oozing of blood. Cut surface showed solid and cystic areas, yellowish- white myxoid areas with old and new haemorrhagic areas. (Figure - 1, B)

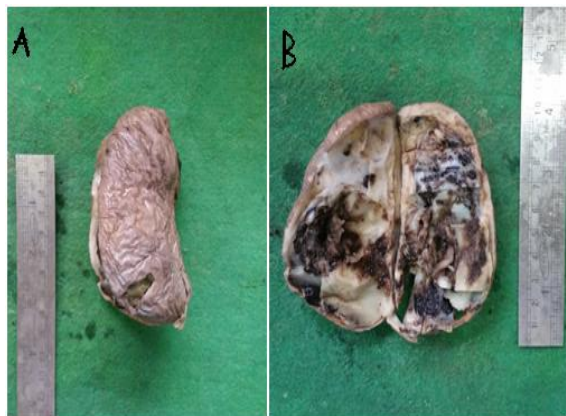


Figure 1: Gross findings- A) External surface, B) Cut-section; solid and cystic areas with old and new hemorrhagic areas.

Microscopic examination: Revealed both Antoni A and Antoni B areas with Verocay bodies (Figure 2). Few cells showed nuclear atypia and other degenerative changes like cyst formation, calcification, haemorrhage and hyalinization (Figure 4). The Schwann cell nuclei appeared to be large and hyperchromatic (Figure 3, A). Immunohistochemistry revealed uniform positivity for S-100 (Figure 3, B). Hence the final diagnosis of Ancient Schwannoma was made.

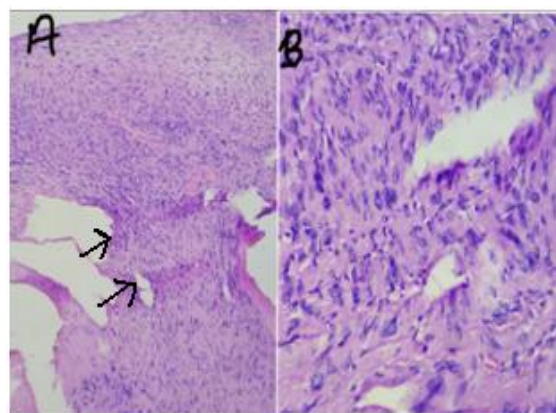


Figure 2: Microscopic areas A) 10x magnification (nuclear palisading {Black arrow}), B) 40x magnification.

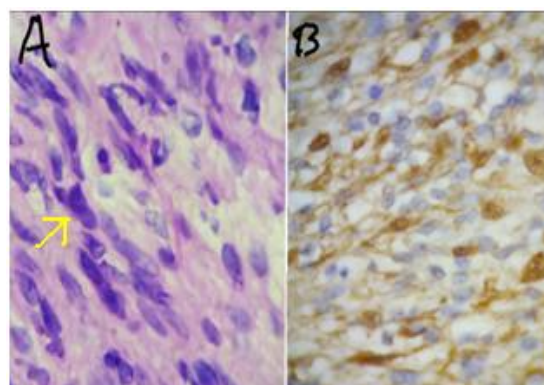


Figure 3: Microscopy A) 100x magnification showing large and hyperchromatic Schwann cell nucleus (yellow arrow), B) S-100 positivity.

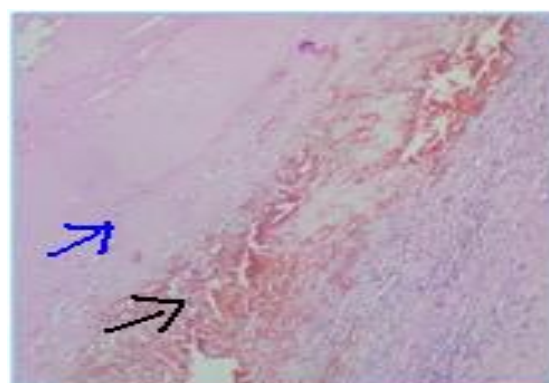


Figure 4: Microscopy 4x magnification showing areas of hemorrhage (black arrow) and hyalinisation (blue arrow).

DISCUSSION

Chest wall schwannoma accounts for 5% of all thoracic neoplasms. ⁽²⁾ It is rarely seen in people below the age of 20 years. ⁽³⁾ Ancient schwannoma constitutes 0.8 % of soft tissue tumors. ⁽⁴⁾ In 1910, Verocay reported a schwannoma as a true neoplasm originating from the Schwann cells which contained no neuroganglion cells. ⁽⁵⁾ In 1935 schwannomas were defined as arising from nerve sheaths and were also known as neuroma, neurilemmoma or perineurofibroblastoma. ⁽⁶⁾ Ancient schwannoma behaves as an ordinary schwannoma; therefore the nuclear atypia can be dismissed as a degenerative change. ⁽¹⁾ Symptoms are usually related to the pressure effects ⁽³⁾ on nearby organs or sensory changes in the distribution of the affected nerve. ⁽¹⁾ Our patient had left-sided chest pain. Complete excision of the swelling was done and sent for histopathological examination. Histopathologically the hallmark of schwannoma is the pattern of alternating

Antoni A & Antoni B areas. The relative amounts of these two components vary. ⁽¹⁾ Antoni A areas are composed of compact spindle cells that usually have twisted nuclei, indistinct cytoplasmic borders and occasionally clear intranuclear vacuoles. They are arranged in short bundles or interlacing fascicles. In highly differentiated Antoni A areas, there may be nuclear palisading, whorling of cells and Verocay bodies, formed by two compact rows of well-aligned nuclei separated by fibrillary cell processes. Mitotic figures are occasionally present but can usually be dismissed if the lesion otherwise has all the hallmarks of schwannoma. ⁽¹⁾ Antoni B areas are far less orderly and less cellular. The large, irregularly spaced vessels, which are characteristic of schwannomas, become most conspicuous in the hypocellular Antoni B areas. ⁽¹⁾ In type B areas the tumor cells are separated by abundant edematous fluid that may form cystic spaces. ⁽⁷⁾ No strict criteria exist for this diagnosis, but I tend to reserve its use for those cases in which either significant nuclear atypia or pleomorphism is seen or in which the cellular (Antoni A) component has become as inconspicuous or focal as to render the diagnosis difficult other than by S-100 staining. ⁽⁹⁾ Our case had similar microscopic features. Differential diagnoses were neurofibroma, perineuroma, dermatofibrosarcoma Protuberans (DFSP). ⁽¹⁾ Neurofibroma & DFSP lack Antoni A & Antoni B pattern of schwannoma. Perineuroma is S-100 negative. The S-100 positivity of neurilemmoma allows distinction from other tumors that exhibit nuclear palisading, such as leiomyoma, and the uncommon neuroblastoma. ⁽⁸⁾ Based on the microscopic features and

positive for S-100, final diagnosis of Ancient Schwannoma was made. Recurrence and malignant transformation is very rare. Our patient is doing very well after surgery.

CONCLUSION

Schwannoma is a common peripheral nerve sheath tumor, which is rare in the chest wall. Clinically and radiologically it can be misdiagnosed as a fibrous tumor. Histopathological examination is essential for confirmation of diagnosis assisted with immunohistochemistry in few cases.

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