

Case Report

Ancient Schwannoma of Vagus Nerve Mimicking Hamartoma

Susmita Kundu¹, Debabani Biswas², Swapnendu Misra³, Shantanu Dutta³ and Annoy Sen³

Department of Pulmonary Medicine, R.G. Kar Medical College¹; College of Medicine and Sagar Dutta Hospital²; and Departments of Respiratory Medicine, Cardiothoracic and Vascular Surgery and Pathology, Institute of Postgraduate Medical Education and Research³, Kolkata (West Bengal), India

Abstract

Schwannomas arising from vagus nerve sheath are rare mediastinal neurogenic tumours. Schwannomas usually arise from left hemithorax. Unlike a hamartoma, radiologically, calcification is rarely seen in schwannomas. We present the rare case of an ancient schwannoma arising from vagus nerve sheath from the right hemithorax presenting with gross calcification.

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Key words: Schwannoma, Popcorn calcification.

Introduction

Different neurogenic tumours, arising from nerve sheath, ganglion cells and paraganglion cells,¹ are commonly seen in the posterior mediastinum. These are usually benign with few exceptions. Neurogenic tumour schwannoma arises from Schwann cells of neural sheath. Majority of schwannomas (45%) occur in the head and neck and only 9% are seen in the mediastinum.² Schwannoma originating in thoracic inlet may arise from any of vagus, sympathetic chain, phrenic or intercostal nerves in the para-vertebral gutter.^{1,3,4} Schwannomas or neurilemmomas arising from vagus nerve sheath are rare mediastinal neurogenic tumours with incidence of only 2% to 4%.^{1,3,5-7} These form a lateral mass alongside the parent nerve. Five different subtypes are described: ancient, glandular, melanotic, cellular, psammomatous melanotic schwannoma.⁸ 'Ancient' term is proposed for neural tumours with long duration, encapsulated, focal calcification, benign, and degenerative changes.⁸

Case Report

A 35-year-old male patient, a manual labourer by occupation, presented with slowly progressive chest pain over anterior chest wall on the right side for last four months. Pain was insidious in onset, dull and boring in nature often radiating to back. The pain was disturbing his sleep, was aggravated by cough, deep inspiration, and jerky body movement and relieved by analgesics only. He had no history of expectoration, shortness of breath, wheeze, haemoptysis or fever. He did not have hypertension, diabetes mellitus. He was a tobacco smoker and regularly consumed alcohol.

General physical examination was unremarkable. Examination of the respiratory system revealed

decreased movements on the right side, a dull percussion note over right Kronig's isthmus, right clavicle, right second and third intercostal spaces along the mid-clavicular line, diminished vesicular breath sounds and vocal resonance in the same regions. Examination of other systems was normal. Chest radiograph (Figure 1) showed a homogeneous opacity over the right upper zone with smooth outline and presence of gross irregular calcifications. Contrast enhanced computed tomography (CT) of thorax (Figure 2) showed a 8.6cm × 7.8cm mass lesion involving the right upper lobe with smooth margin, and areas of low attenuation [28 Hounsfield units (HU)] interspersed with areas of popcorn-like calcification (474 HU). There were no bony erosions, involvement of mediastinal structures, pleura or displacement of great vessels.

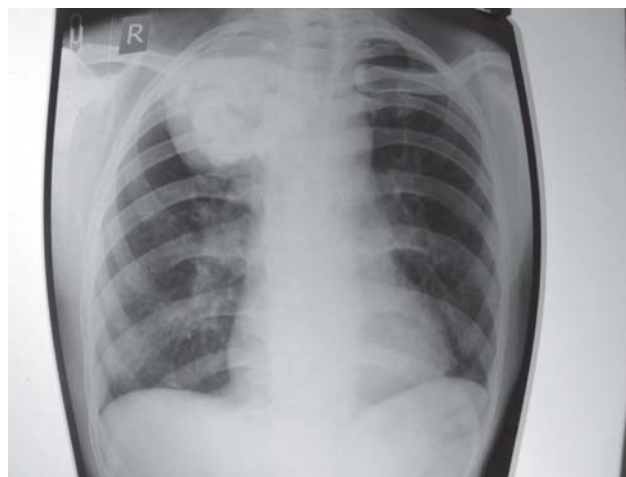


Figure 1. Chest radiograph (postero-anterior view) showing a mass in right upper zone with dense irregular calcification.

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Correspondence and reprint requests: Dr Susmita Kundu, Flat 101, Monomohini Apartment, 71, Shyamnagar Road, Kolkata-700 055 (West Bengal), India; Phone: 91-9433238525; E-mail: susmitakundu.chest@yahoo.com

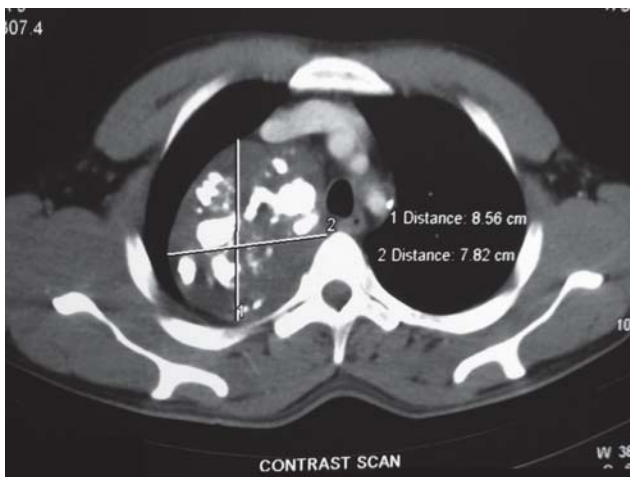


Figure 2. Contrast enhanced CT of thorax showing right upper lobar mass with calcification.

Ultrasonography-guided tru-cut biopsy of the mass was done and histopathological examination showed linear strips of tissue showing spindle-shaped cells lying in a myxoid background. Contrast enhanced CT of brain was normal. Haematological and biochemical investigations were within normal limits. Abdominal ultrasonography was within normal limits also. The patient was diagnosed to have a possible huge hamartoma with popcorn calcification involving right upper lobe of lung. He underwent right postero-lateral thoracotomy. A hard nodular circumscribed mass was found arising from upper part of right chest cavity attached peripherally to the vagus nerve and pushing the lung down. The mass was extending into the thoracic inlet and getting vascular supply from subclavian artery. Macroscopically it was a heterogeneous mass with gross white areas. Histopathological examination revealed a tumour mass composed of mainly spindle- to oval-shaped cells in a

myxoid background suggestive of schwannoma (Figures 3A and 3B). Mitotic figures were absent and focal areas of calcification noted.

Alternate areas of hypercellular (Antoni A) and hypocellular (Antoni B) were also seen. Immunohistochemistry was strongly and diffusely positive for S100 and negative for Desmin and smooth muscle actin (SMA) confirming the diagnosis of ancient schwannoma. The six-month post-operative follow-up was uneventful.

Discussion

The present case represents the rare occurrence of a huge ancient schwannoma involving the inlet of right hemithorax and pushing the lung apex down in a young adult. Schwannomas are equally seen in both genders in their third and fourth decades of life.¹ It is mostly asymptomatic, encapsulated, lobulated with smooth margin and remains peripherally attached to at least one nerve root. Schwannoma of vagus is more commonly reported on the left side compared to the right, as seen in the present case.^{3,9} Complete surgical resection is usually possible sparing the parent nerve as the tumour is generally separable from the underlying structure. Radiologically, heterogeneous appearance with degenerations and cystic appearance is more common in schwannoma than neurofibroma, another common benign peripheral nerve sheath tumour. "Ancient" schwannoma refers to long standing tumour with advanced degenerative changes including calcification, hyalinisation and cavitations.⁸ Tumoural popcorn calcification is usually seen in chondroid lesions (e.g., enchondroma, chondroma), fibrous dysplasia, pulmonary hamartoma, granuloma, and degenerating fibroadenoma of breast.¹⁰ Popcorn calcification in a solitary, well-defined pulmonary nodule is virtually diagnostic of pulmonary

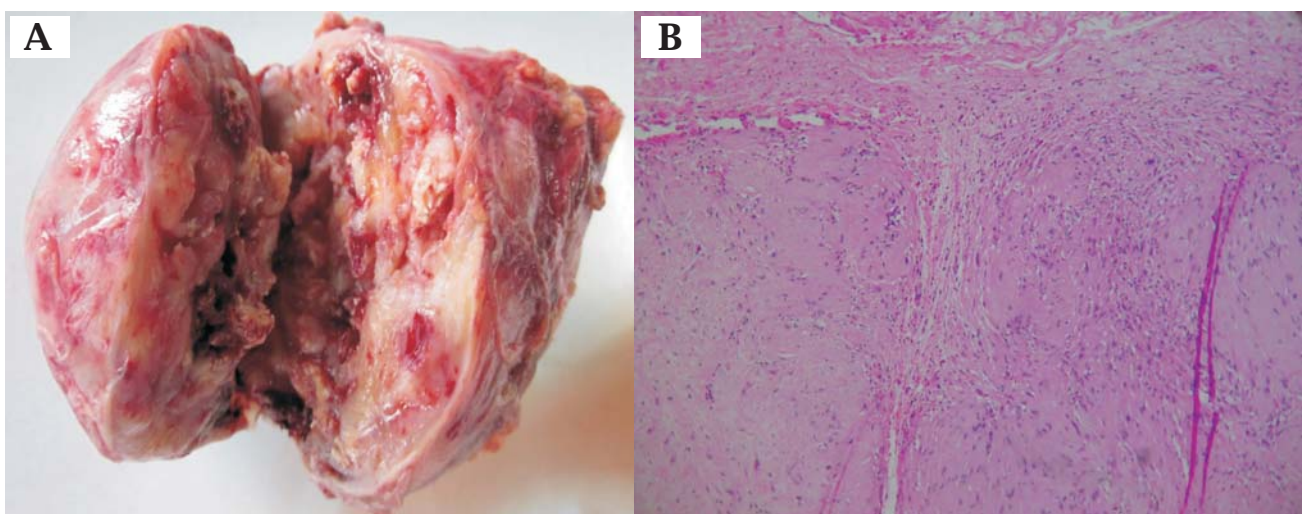


Figure 3. (A) Macroscopic specimen of the resected mass measuring 15cm×10cm and (B) photomicrograph showing alternate hypercellular (Antoni A) and hypocellular (Antoni B) areas with foci of calcification and absent mitotic figures (Haematoxylin and eosin × 100).

hamartoma. Frequency of calcification increases to more than 75% in lesion more than 5cm. Presence of fat in a well-circumscribed nodule may be due to pulmonary hamartoma, lipoma, myolipoma, and metastatic disease. Fat can be recognised by comparing it to subcutaneous fat that has an attenuation of -40 HU to 120HU.

However, in the present case, because of the presence of low-attenuated areas resembling fat planes interspersed with extensive popcorn calcification in a huge mass with smooth outline the lesion was confused with a pulmonary hamartoma. Later diagnosis of ancient schwannoma was confirmed by the histopathological examination and immunohistochemistry study of the resected sample. *To the best of our knowledge*, though calcification is often present in ancient schwannomas, the extensive popcorn-like calcification in schwannoma of vagus nerve has not been reported so far. The purpose of reporting the case is to highlight the rare radiological finding of popcorn calcification in a schwannoma originating from right vagus nerve at thoracic inlet.

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