

Neurofibroma Arising from Phrenic Nerve

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ABSTRACT

We report a case of neurofibroma arising from the left phrenic nerve and not associated with neurofibromatosis in a 46-year-old male. The patient presented with gradually progressive breathlessness and pain in chest for six months. Radiological investigations revealed a posterior mediastinal mass on the left side of the chest. On postero-lateral thoracotomy, the resected tumour was found to be arising from the phrenic nerve. Histopathological examination confirmed it to be a neurofibroma. Absence of any other distinctive lesions of neurofibromatosis makes this an unusual presentation. [Indian J Chest Dis Allied Sci 2012;54:263-264]

Key words: Neurofibroma, Phrenic nerve, Neurofibromatosis, Mediastinal mass.

INTRODUCTION

Neurogenic tumours constitute a large population of mediastinal tumours, accounting for 10% to 20% of all mediastinal masses.^{1,2} The posterior mediastinum is the most common site to be involved. These arise most commonly from intercostal, or sympathetic nerves and occasionally, from the vagus nerve.³ A neurofibroma, a benign nerve sheath tumour of the peripheral nervous system, arising from the phrenic nerve is a very rare entity. Till date only two cases have been reported.^{2,4} In 2004 Saito *et al*² reported the first case of mediastinal neurofibroma originating from the phrenic nerve in a patient without von-Recklinghausen disease neurofibromatosis. Mohamed and Colleagues⁴ reported another case recently. We report a rare case of neurofibroma arising from the phrenic nerve which was not associated with neurofibromatosis.

CASE REPORT

A 46-year-old middle-aged man presented with complaints of gradually increasing breathlessness and a dull aching pain on the left side of the chest since six months. He did not have a history of fever, cough, haemoptysis, and exposure to carcinogenic materials, or any other illness and was not a known alcoholic or a smoker. The family history did not reveal any hereditary lung diseases.

The general physical examination was unremarkable. On examination of the respiratory system, the left thoracic region was slightly prominent. The left hemithorax was 4cm larger than the right hemithorax. There was no tenderness on

palpation. Percussion notes were impaired all over the left lung fields. Breath sounds were reduced in intensity on the left inter-scapular, supra-scapular and infra-scapular areas. Examinations of other systems revealed no abnormality.

The chest radiograph (postero-anterior view) showed a homogeneous well-circumscribed opacity in the left upper and mid zones of the chest (Figure 1). The trachea was pushed to the opposite side. The right lung field showed a patch of consolidation in the lower zone, while remaining lung fields were



Figure 1. Chest radiograph (postero-anterior view) showing a homogeneous, well-circumscribed opacity in the left upper and mid zones of the chest.

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clear. Cardiac size was within normal limits. Bony cage and soft tissues were normal. A posterior mediastinal mass with right lower zone pneumonitis was the provisional diagnosis.

Computed axial tomography (Figure 2) confirmed a heterodense, well-defined posterior mediastinal mass occupying most of the left hemithorax. No mediastinal lymphadenopathy was noted. It also showed patch of consolidation and thickened pleura/minimal effusion.

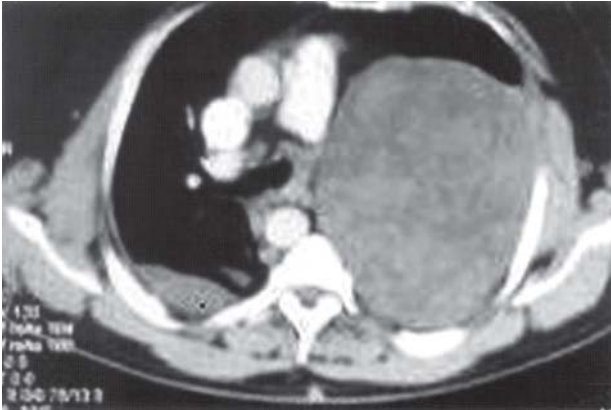


Figure 2. Computed axial tomography showing heterodense, posterior mediastinal mass occupying most of the left hemithorax. No mediastinal lymphadenopathy was noted. It also showed patch of consolidation and thickened pleura/minimal effusion.

The tumour was operated by a left postero-lateral thoracotomy under general anaesthesia after consent and pre-operative evaluation. It was seen to arise from the left phrenic nerve, spheroidal in shape, measuring 14.5cmx8cm and was well-encapsulated.

Histopathology examination (Figure 3) revealed spindle cells arranged in fascicles. The nuclei were elongated with wavy, serpentine configuration and had pointed ends with moderate amount of eosinophilic cytoplasm. Blood vessels showed hyalinisation. Features were consistent with a neurofibroma. There

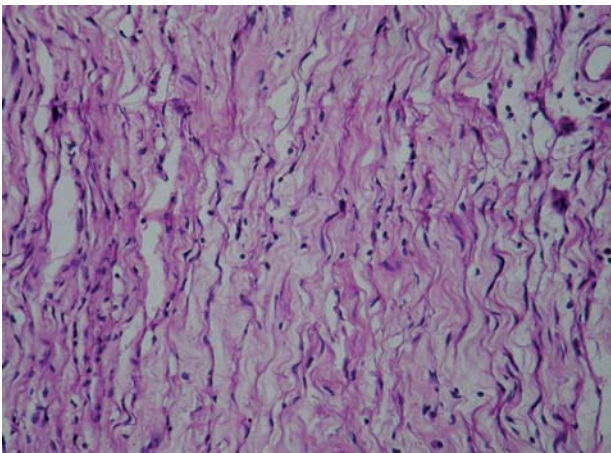


Figure 3. Histopathology of the tumour. Spindle cells with elongated nuclei are seen arranged in fascicles.

were no post-operative complications and the patient recovered gradually. Recurrence has not been reported after one year follow-up period.

DISCUSSION

Neurofibromas of the mediastinum constitute 11% of all mediastinal masses.¹ Usually neurofibromas arise in association with Von-Recklinghausen's disease.⁵ It is interesting to note that our case did not have *café-au-lait* spots, or optic glioma or any other distinctive lesions of neurofibromatosis. Thoracic manifestations of neurofibromatosis that have been published as case reports include ground-glass opacities, bibasilar reticular opacities, bullae, cysts and emphysema of lung,⁶ spinal deformities leading to kyphoscoliosis and consequent respiratory impairment,⁷ diffuse interstitial lung diseases,⁸ and pulmonary hypertension.⁹ In our case, however, none of these features were noted.

To the best of our knowledge, only two cases of a neurofibroma arising from the phrenic nerve and not associated with neurofibromatosis have been reported so far.^{2,4} Our case is consistent with the findings in those cases.

The lack of specific symptomatology and radiologic features make pre-operative diagnosis extremely difficult. However, the location, i.e. the posterior compartment of the mediastinum is suggestive of the diagnosis. It is important to note that this condition is potentially treatable.

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