A case of pleural neurofibroma: A rare pleural tumor

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ABSTRACT

Neurofibromas (NFs) are benign tumors involving any nerve from the root level to the smallest branch. They may enlarge locally and cause a pressure effect on thoracic structures such as trachea, superior vena cava, and esophagus. Pulmonary and pleural involvement is very rarely seen. We have reported a case of a pleural effusion secondary to lymphatic obstruction due to remnant NF for which patient had undergone surgical resection in the past.

Key words: *Dyspnea, Neurofibroma, Tumor*

Pleural effusion is caused due to infections, neoplasms, trauma, drugs, collagen vascular disease, iatrogenic, or it can be idiopathic when no evident cause is available [1]. Neurofibromas (NFs) are benign tumors arising from neural crest cells. They can involve any peripheral nerve. Neurogenic tumors account for about 9% of primary mediastinal masses in adults and 30% of mediastinal tumors in children [2]. NF rarely involves the pleura, and to the current knowledge of author, no such case has been reported so far.

CASE REPORT

A 31-year-old male presented to hospital with the complaints of dyspnea and left-sided chest pain for a month. There was no history of fever, weight loss, cough, or loss of appetite. On general physical examination, he was of average built with no peripheral lymph nodes palpable. His oxygen saturation at room air was 95% with a pulse rate of 95/min. On system-specific examination, breath sounds were absent on auscultation on the left side of the chest in inframammary, infra-axillary, and infrascapular area. He was a non-smoker with no prior history of antitubercular treatment. Chest X-ray showed a massive left pleural effusion (Fig. 1). Ultrasonography (USG) chest also confirmed it to be effusion. The patient's history revealed that the patient had undergone an operation for a posterior mediastinal cystic lesion about 6 months back (Fig. 2), and the histopathology was consistent with NF.

Routine blood investigations were normal. A computed tomography (CT) scan of the chest revealed only the left pleural effusion with passive collapse of the underlying lung with no mass lesion and normal mediastinum (Fig. 3) Pleural fluid was analyzed, and it was transudative in nature with no atypical or malignant cells. Triglycerides in pleural fluid were also within normal limits. USG of the abdomen and all other routine investigations were within normal limits. There was no involvement of skin, bone, and peripheral nerves.

Video-assisted thoracoscopic pleural biopsy was done, and intercostal tube was inserted into the left pleural cavity. Histopathology showed it to be a NF with immunohistochemistry showing s-100 positivity (Fig. 4). The ICD was removed when the discharge from the pleural cavity was minimal, and the patient was followed with chest X-rays to look for any refilling on an outpatient basis. There was no recollection of fluid, and the patient was clinically well in the follow-up. (Fig. 5).

DISCUSSION

Due to the vast distribution of peripheral nerves throughout the thorax, NFs may involve the ribs, chest wall, lungs, and mediastinum. Classic imaging findings include small, well-defined subcutaneous NFs, focal thoracic scoliosis, posterior vertebral scalloping, enlarged neural foramina, and characteristic rib abnormalities due to bone dysplasia or erosion from adjacent NFs. Neurogenic tumors account for about 9% of primary mediastinal masses in adults and 30% of mediastinal tumors in children [1]. NFs are benign tumors, which may involve any nerve from the root level to the smallest branch. They may enlarge locally and cause pressure effects on thoracic structures such as trachea, superior vena cava, and esophagus. Malignant transformation is seen only in cases associated with neurofibromatosis type 1; however, very little is known about the biological events involved in the malignant transformation of NFs [3].

Pulmonary and pleural involvement is very rarely seen. Pulmonary involvement may lead to chest deformities, airway and parenchymal neurogenic tumors, pulmonary fibrosis, cystic pulmonary diseases, primary pulmonary hypertension, central hypoventilation, and diaphragm paralysis [4]. Pleural involvement can be direct when these tumors arise from the intercostal

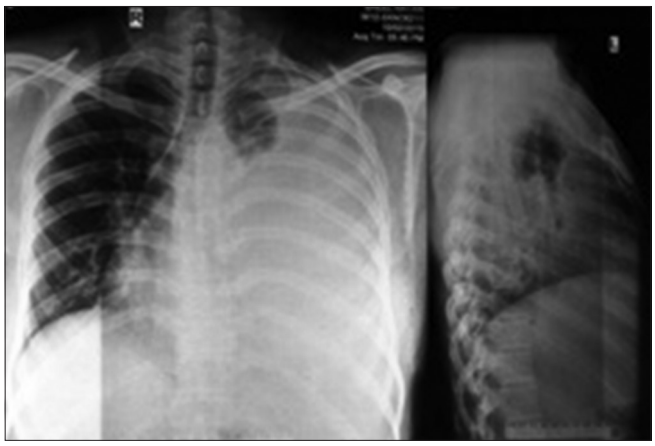

Figure 1: Chest X-ray posterior-anterior and lateral view showing the left pleural effusion

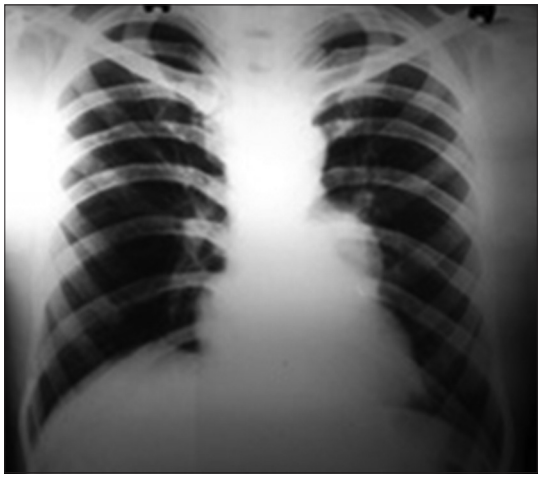

Figure 2: Previous X-ray (6 months back) showing the left hilar shadow

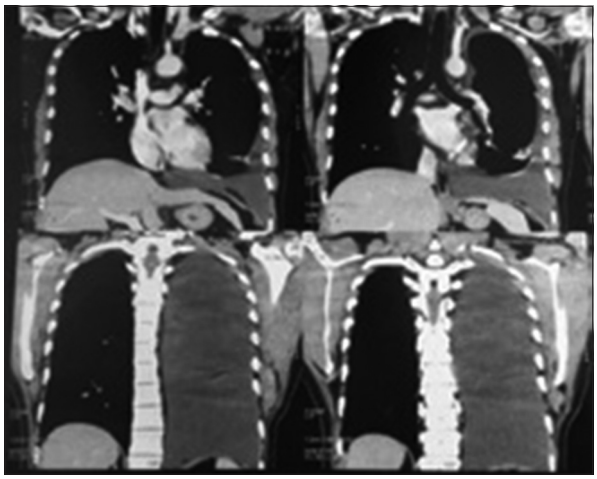

Figure 3: Computed tomography chest showing the left effusion with no residual mass in posterior mediastinum

nerves. They can also involve the pleura indirectly by causing lymphatic obstruction and hence leading to pleural effusion. The patient usually is presented with chest pain on the affected side and dyspnea due to effusion. Two cases have been reported by Langman and coll presenting with shoulder pain and no chest-related symptoms [5].

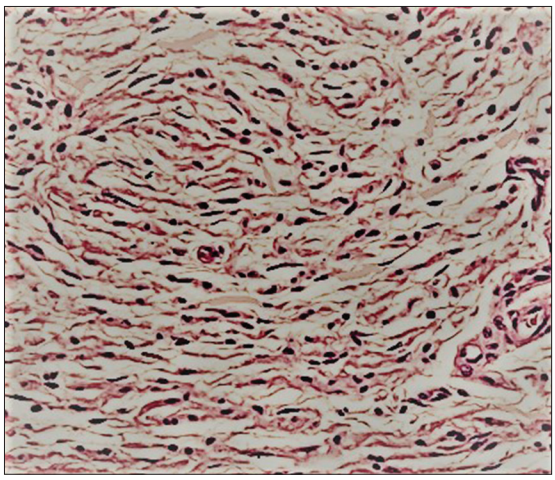

Figure 4: Histopathological slide shows neurofibroma with s-100 positivity

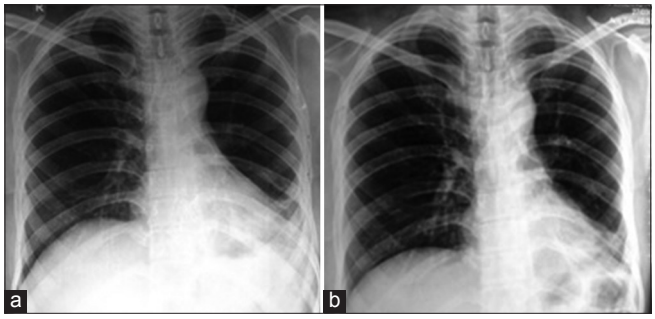

Figure 5: Chest X-ray after (a) 1 month and (b) 2 months of surgical drainage of fluid shows the left pleural thickening with no recurrence of effusion

Our patient presented with dyspnea and effusion. He was operated in the past for posterior mediastinal NF. CT chest did not reveal any pleural nodules or pleural-based mass lesion, which is a characteristic feature of primary pleural NF arising from intercostal nerves. In this case, pleural involvement was probably due to the lymphatic spread of old remnant NF and effusion because of lymphatic obstruction. The patient was asked for a regular follow-up so that timely intervention can be done if there was any recurrence of effusion.

CONCLUSION

The treatment of NF depends on the size and the location of the tumors and should be conservative. Surgery should be the option if the tumor involves the adjacent organs. Surgical resection may be incomplete in mediastinal NFs, and hence, mediastinum can be irradiated to prevent recurrence postsurgery.

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