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Desmoid fibromatosis of lung: a rare thoracic neoplasm

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ABSTRACT

Desmoid fibromatosis is a very rare neoplasm. Intrathoracic desmoid tumour is even rarer subset of this group and only a handful of cases has been reported in medical literature till now. We here report the case of a 34 year old female from Eastern India who presented with refractory pain in the left axilla and inner arm. Thoracic imaging revealed a large mass in left lung apex surrounding the great vessels and brachial plexus nerves. The mass was completely excised surgically and histopathology revealed spindle-shaped cells in a collagenous background. Beta-catenin was strongly positive and SMA was focal positive. The patient was not given any radiotherapy. Intrathoracic desmoid tumour is extremely rare but timely treatment can be rewarding. Immuno histo chemistry (IHC) plays a significant role in its diagnosis. Clinicians should be aware of this rare neoplasm because it usually requires urgent surgery. The patient should be followed up for local recurrence.

INTRODUCTION

Desmoid tumour is a proliferative disorder of fibrous connective tissue that can range from benign soft tissue mass to aggressive fibrosarcoma [1]. According to the WHO definition, these are tumours with a high chance of recurrence but very low probability of metastasis [1].Desmoid, in general, is a very rare neoplasm with projected incidence of 3-4 per million [1].

Desmoid tumours are mainly found in the abdomen, retroperitoneum or thoracic wall [2]. Intrathoracic desmoid tumour is an extremely rare neoplasm with only a few cases found in medical literature [3]. We here report a case of desmoid tumour of lung from Eastern India.

THE CASE REPORT

A 34 year old female from West Bengal presented with gradually progressive pain in the inner side of left arm for three months. The pain was ill-localized, burning in character and often disturbed sleep at night. She also complained of mild decrease in grip strength of the left hand and heaviness around left shoulder. There was no history of trauma. There was no other clinical symptom, no history of weight loss. The patient had been treated with various analgesics with no effect.

At presentation to our institution, the patient was clinically stable. She complained of nagging burning pain around left axilla and inner side of left arm. But no definite sensory loss was demonstrable. There was no muscle wasting. Trachea was in the midline. Examination of the thorax revealed absent breath sound in left apex with dull percussion note. There was no lymphadenopathy or rib tenderness. She did not have cough or change of voice.

Initial laboratory tests revealed hemoglobin of 11 g/dl, total Leukocyte count of 8000/µL (normal differentials) and platelet count of 160 000/µL. ESR was 50 mm in the 1st hour. Urea/creatinine/electrolytes and liver function tests were all normal. A digital chest X ray showed (figure 1) a large well-circumscribed mass in the left apex with no rib involvement or pleural effusion. A CECT scan of thorax was done (figure 1) which showed a large mass in left apex infiltrating the great vessels and nerve fibres. There was no radiological evidence of rib invasion. CT guided biopsy from the mass was done which showed a fibrocollagenous stroma with spindle shaped cells.

The patient was immediately transferred to the cardiothoracic surgery department where complete excision of the mass was done (figure 2). Intraoperatively, the mass was found to encase, but not invade, the great vessels of superior mediastinum and

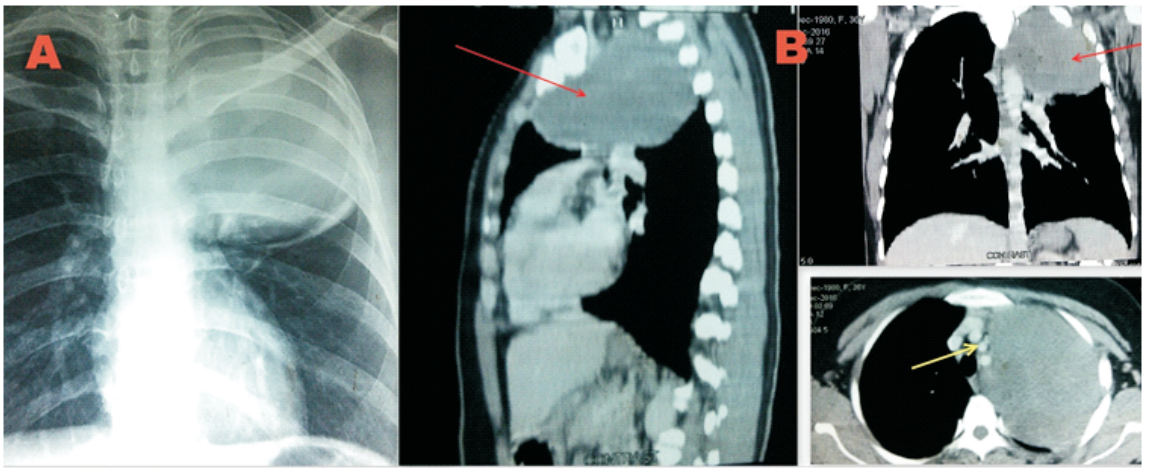

Figure 1: A: chest x ray showing a large mass in left lung apex; **B:** CECT thorax showing the large mass (red arrow) infiltrating blood vessels (yellow arrow) and nerves

posterior cord of brachial plexus. However, exact site of origin could not be found. Biopsy from the mass showed interlacing fascicles of spindle-shaped cells admixed with collagen (figure 3). There were no mitotic figures or nuclear atypia. Immuno histochemistry was done from the mass which showed (figure 4) positive beta-catenin, focal positive SMA and negative for CD34, CD117, ALK-1 and Desmin. Thus, the tumour was finally diagnosed as desmoid fibromatosis of lung. Her post-operative course was uneventful. At 6 and 12 months' follow up, the patient had not had any radiological or clinical sign of local recurrence. A follow up colonoscopy was also done, but it did not show any evidence of Gardner's syndrome. A mild burning pain in the left axilla was persisting at last visit.

DISCUSSION

Intrathoracic Desmoid tumour is an extremely rare neoplasm. Most reported thoracic desmoid tumours are actually chest wall neoplasms with invasion of lung [3]. But sometimes, true pleural or pulmonary masses have been reported [4]. As of 2009, only 26 cases of desmoid tumour of chest has been reported in English literature [5]. Because of the rarity, the clinical presentation of the tumour is still not known with certainty. However, past reports have shown presentation with chest pain, dyspnoea or mild cough [3, 5]. Desmoid tumours have been sometimes reported to develop in areas with previous trauma or surgery [6]. But our patient had no such history.

The exact aetiology of desmoid tumours is not known. Different hypotheses have proposed genetic factors or an abnormal reaction to previous local trauma as probable aetiologies [5]. The tumour is commonly found in 2nd and 3rd decades of life, especially in females [1]. Thus some authors have also proposed a hormonal factor (like estrogen) in its causation [1, 5].

Thoracic desmoid tumour may mimic other similar soft tissue neoplasms. Hence, immunohistochemistry is needed for confirmation. Beta-catenin expression is an important marker for diagnosis of pleuropulmonary desmoid [7]. This was strongly

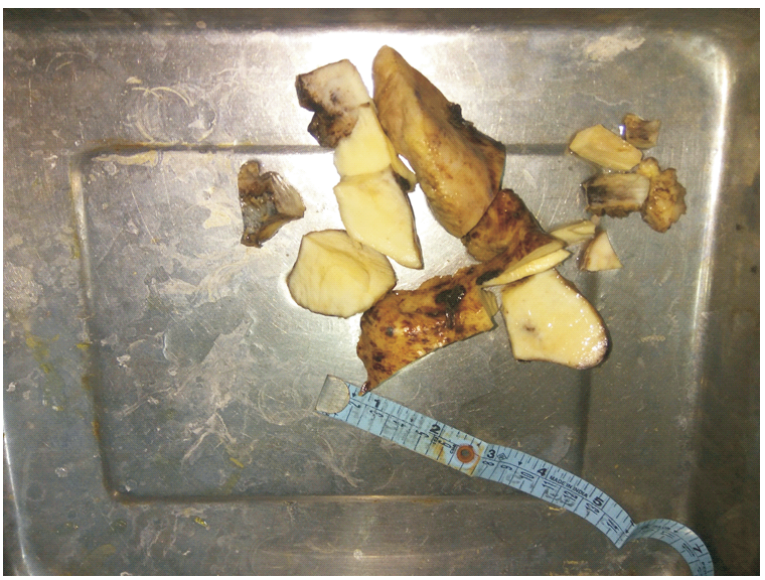

Figure 2: The operated specimen

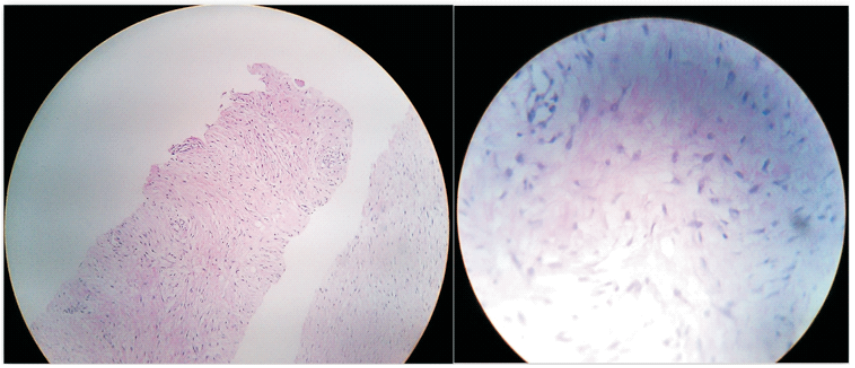

Figure 3: Microscopic picture of the mass showing spindle cells in a collagenous background

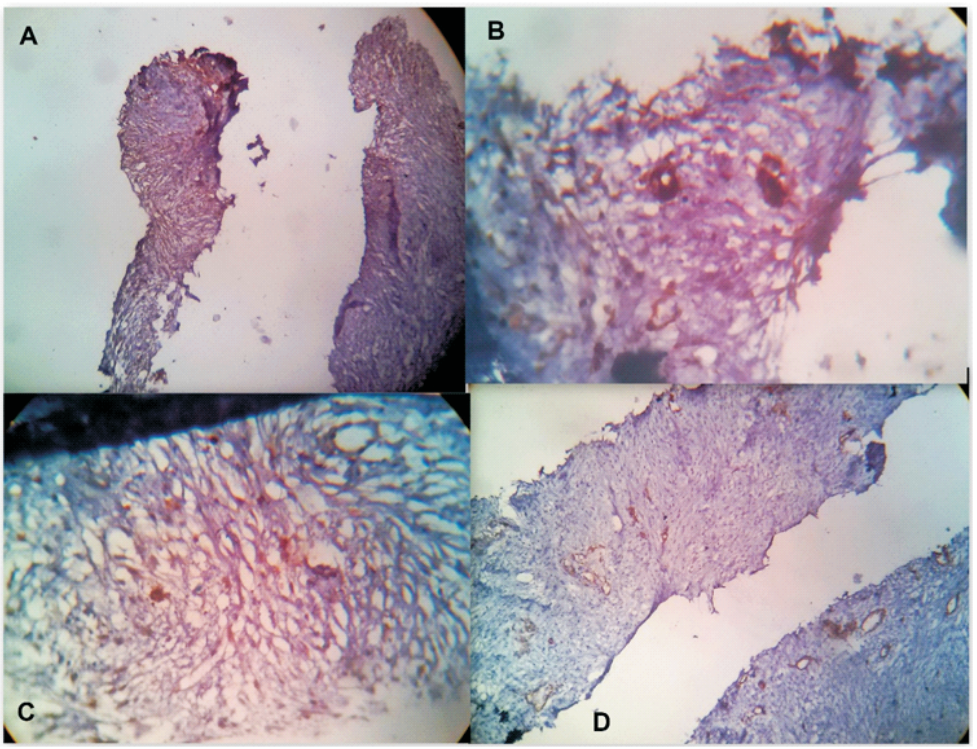

Figure 4: Immunohistochemistry of the mass showing focal positivity of SMA (A), Desmin negativity (B), CD117 negativity (C) and CD34negativity (D)

positive in our patient. Desmin, CD34, ALK-1 and pancytokeratin are usually negative. Desmin may be positive in some cases [7]. In our case, the tumour was negative for Desmin. In the gross specimen, desmoids usually show a white or tan appearance, as in figure 3. Presence of areas of haemorrhage or necrosis suggests malignant transformation [8].

Treatment of thoracic desmoid is a contentious issue. As the number of reported cases is very small, no standard protocol is there. Some of the reported cases have shown good result with only surgery [3]. Others have reported successful therapy with a combined modality of surgery and radiotherapy. However, chance of recurrence is quite high and regular follow up is needed [5]. Usually recurrence occurs locally. Only rarely, desmoid tumours elsewhere is the body have been seen to recur in the lungs [9]. Recurrence is usually treated in the same way as the primary tumour. If there are multiple sites of recurrence, surgery may not be feasible and then, only follow up is recommended [9]. However, despite the high chance of local recurrence, survival data for this tumour is quite encouraging. Data for chest wall desmoid have shown a 5-year survival of more than 90% [10]. Hence, early treatment can be quite rewarding.

CONCLUSION

The present case is unique because of the rarity of the tumour and also its location. Clinicians should be aware of rare thoracic neoplasms. Early diagnosis and proper treatment can substantially improve the prognosis.

Abbreviations: WHO: World Health Organization; ESR: Erythrocyte sedimentation Rate; CECT: Contrast enhanced CT scan; SMA: Smooth muscle actin

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