An unusual case of chest wall desmoid tumor

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Abstract Desmoid tumor of the chest wall are uncommon fibromatous tumors characterized by their local invasion and frequent recurrences. Extra-abdominal sites are mainly the shoulder girdle, the pelvic girdle and distal ends of the lower limbs. The chest wall represents 8–10% of cases and the tumor is exceptionally intrathoracic. We present here a rare case of a large desmoid tumor of left antero-lateral chest wall. In the presented case, wide excision of the tumor and chest wall reconstruction was done. There was no recurrence in 3 years of follow up.

Keywords Extra abdominal tumor · Chest wall · Surgery

Introduction

Desmoid tumors are rare connective tissue tumors. They account for approximately 3.5% of fibrous tumors, 0.3% of all solid tumors and only 0.03% of all the neoplasms [1–3]. Desmoid tumors of the chest wall are uncommon fibromatous tumors characterized by their local invasion and frequent recurrences. These tumors are usually asymptomatic and painless but can be painful due to nerve involvement in the form of entrapment or compression. [1, 4]. We report a case which presented with complaint of chest pain and intermittent fever since 2–3 years.

Case report

A 50-year-old-male patient was referred to us with complaints of chest pain on left side and fever on and off. Patient had similar episodes of chest pain since 2–3 years and was taking conservative treatment but did not improve. Patient had no other complaints pertaining to respiratory system i.e., dyspnea or cough. Patient was a chronic smoker.

On physical examination there was a palpable mass 10 × 9 cm on left anterolateral chest wall. There was dullness along the left mammary region. Chest X-ray revealed homogenous opacity in the left lateral wall of chest with obscured rib margins. Blood investigations were within normal limits. Computed tomography (CT) scan revealed a homogenous mass of soft tissue density, measuring 8.35 × 5.36 cm, arising from left anterolateral wall of thoracic cage with adjoining rib destruction (Fig. 1). Specks of calcification were seen within the mass. Parietal pleura was involved. Paraseptal emphysema was present in the chest wall. Rest of the lung parenchyma was normal. Provisional diagnosis of fibroma with malignant potential was made based on CT findings.

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Fig. 1 Contrast enhanced computed tomography (CECT) scan revealed a homogenous soft tissue density pleural based mass with adjoining rib destruction and rest of lung was normal

Fine needle aspiration cytology (FNAC) was done to obtain tissue diagnosis and it came out as spindle cell tumor.

Patient was taken for operation and left thoracotomy was done with excision of tumor along with resection of 4th, 5th and 6th ribs. Intraoperative findings were noted meticulously. There were 2 tumors in left side of thoracic cage. The bigger one was measuring 10×12 cm (Fig. 2)

Fig. 2 Gross specimen of resected tumor

and the smaller one was of dimension 3×4 cm in size. Major portions of the tumors were arising from the 4th and 5th rib. Visceral pleura were intact and uninvolved. Parietal pleura was adherent to the tumor mass but was not infiltrated. Tumor was not invading lung tissue. Rest of the lung was found normal. The defect was closed with mesh and wiring to reconstruct the thoracic cage.

Histopathologically tumor showed a hypocellular arrangement of spindle shaped cells with a fibromyxoid background hence the diagnosis of fibromatous desmoid tumor was made. There was no recurrence in 3 years follow up.

Discussion

John Macfarlane first described desmoid tumor in 1832 [3, 5] and Muller in 1838 coined the term “desmoid” from Greek words that mean “tendon-like” [6]. They are rare tumors which were given different names: Desmoid fibroma, aggressive fibromatosis, desmomas and desmoplastic fibroma [3] and currently being referred as the sarcoma of low grade malignancy [7]. The term aggressive fibromatosis is preferred due to its tendency to local invasion and frequent recurrences even after complete surgical resection. These tumors can cause significant morbidity and mortality by their propensity for local invasion and recurrence following surgical excision. Desmoid tumors arise from fascia and connective tissue of the muscular layer and invade surrounding structures. These tumors usually occur more in females than in males in age group of approximately 18–30 years. Most of the patients are asymptomatic. Here we are reporting a rare case of large desmoid tumor with symptoms in male patient. Desmoid tumors are usually non-inflammatory asymptomatic masses. They become symptomatic when the mass itself starts compressing surrounding structures especially nerves. Definitive diagnosis requires histopathological examination supplemented with chest X-ray, CT and MRI of the affected system.

Wide resection of the involved soft tissues and bony structures is the best treatment. Radiation therapy is usually used when a local excision is not feasible. Extra-abdominal desmoid tumors have a high local recurrence rate after resection. In a recent series of chest wall desmoid tumors although the overall 5 years survival rate was 93%, the 5 years recurrence rate despite aggressive surgical excision was 29% [1]. The follow up of desmoid tumors must be done meticulously and for longer duration. Late recurrences can occur even in completely excised lesions, as the residual tumor may be in relation with some permeation node [3]. In our case, we have follow up of 3 years and the patient is not having any recurrence.

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