


Desmoid fibromatosis of the chest wall

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Introduction

Desmoid tumours (also called aggressive fibromatosis) are benign myofibroblastic neoplasms originating from muscle aponeurosis. These tumours are characterized by the proliferation of fibroblast- and myofibroblast-type spindle cells [1]. These tumours are most commonly located in the abdomen. Desmoid fibromatosis of the chest wall is a rare condition.

Case Report

A 70-year-old woman presented with a 12-month history of persistent pain in the right shoulder blade and paresthesia over the right upper breast. She underwent hookwire excision of a right breast lump in 2010, which showed fibrocystic disease with no dysplasia or malignancy. There was no other significant medical or trauma history. Examination of the right shoulder and chest wall was normal. A chest X-ray showed a 5 cm circumscribed mass in the right apex (Fig. 1A). A subsequent chest computed tomography (CT) scan confirmed a well-delineated, low-density 5 cm lesion arising from the chest wall in the right anterior upper lobe with abutment of the pleura with obtuse angle (Fig. 1B). A CT-guided biopsy demonstrated cores of tissue composed of spindle cells with eosinophilic cytoplasm and

Abstract

We report a case of desmoid fibromatosis of the chest wall. A 70-year-old woman was referred to our hospital with right shoulder blade pain and paresthesia over the right upper breast. Chest X-ray and computed tomography demonstrated a 5 cm right apical mass in the chest. Biopsy of the mass demonstrated features of desmoid fibromatosis. The patient subsequently underwent surgical resection of the mass and received adjuvant radiation therapy for microscopic positive margins. In conclusion, although desmoid tumour of the chest is rare, it is worth considering in the differential diagnoses of chest wall tumours.

oval to elongated nuclei (Fig. 2). Focal staining was positive with Beta Catenin, Smooth Muscle Actin, S100 Protein, and Desmin. The specimen tested negative for CD117, Calretinin, Melan A, DOG1, CD34, and AE1/AE3/CK8/18.

Magnetic resonance imaging (MRI) demonstrated a T1 isointense circumscribed mass and T2 markedly hyperintense and avidly heterogeneously enhancement in the right hemithorax apical region. There was no significant enlargement in the 6-week interval period. The mass was bulging into the first intercostal space and had a very broad site of direct abutment to the chest wall. There was no evidence of marrow infiltration. There was no abutment to the brachial plexus although they were in close proximity (Fig. 1C, D).

A right thoracotomy revealed a dusky, well-circumscribed mass with a smooth surface originating at the chest wall apex, with infiltration into the second rib (Fig. 3). The first rib was free from tumour. The mass was excised with a segment of involved second rib. The lung was completely free from the mass. Immunostains confirmed previous biopsy findings. Histology showed features consistent with desmoid fibromatosis arising from the chest wall with extension into the skeletal muscle. The patient was referred to radiation oncology for adjuvant radiation therapy in view of microscopic positive margins. She remains well 3 months after the operation.

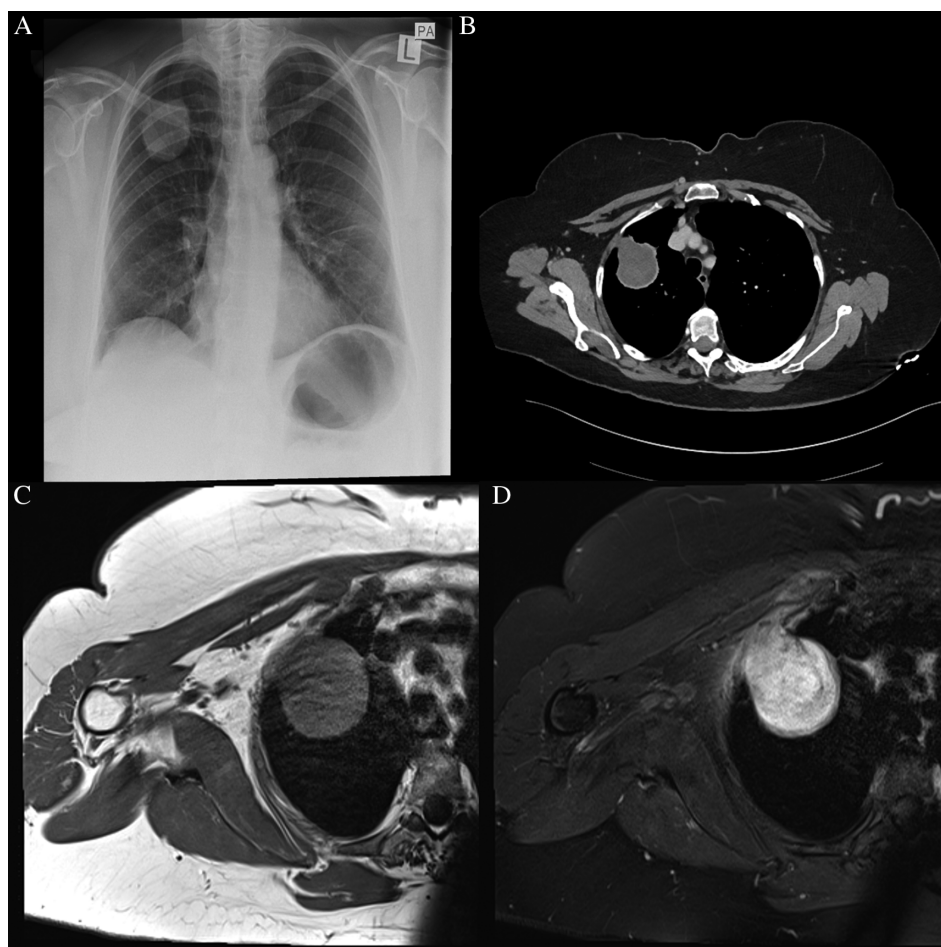


Figure 1. Chest X-ray demonstrating circumscribed mass in the right apex (A). Chest computed tomography (CT) reveals 5 cm lesion in the right anterior upper lobe (B). Magnetic resonance imaging (MRI) axial T1 demonstrating isointense lesion measuring 5.9 × 5.1 × 5.1 cm (C) and hyperintense lesion with band-like columns of low signal intensity on axial T2 (D).

Discussion

The incidence of desmoid tumours is reported to be around 5–6 per million population per year [2]. In 8–10%

of cases, these are located in the chest [3]. When chest involvement is present, these are known to be locally aggressive with a high recurrence rate [4]. These tumours are thought to be primary or secondary to trauma, including surgical procedures, familial adenomatous polyposis,

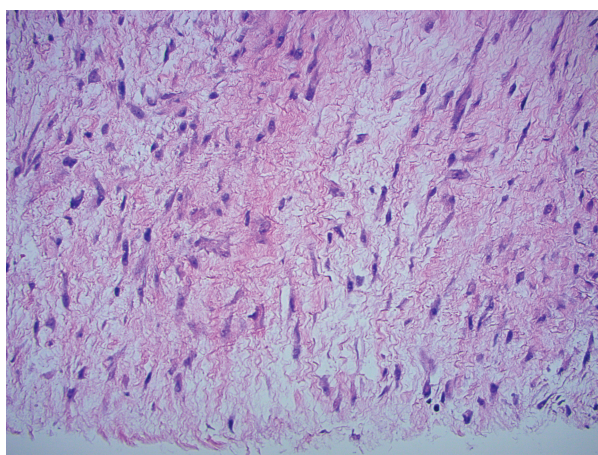


Figure 2. Haematoxylin and eosin (H&E) biopsy specimen 40x. Spindle cells with oval to elongated nuclei observed. No evidence of pleomorphism, mitotic activity, necrosis, or haemorrhage.

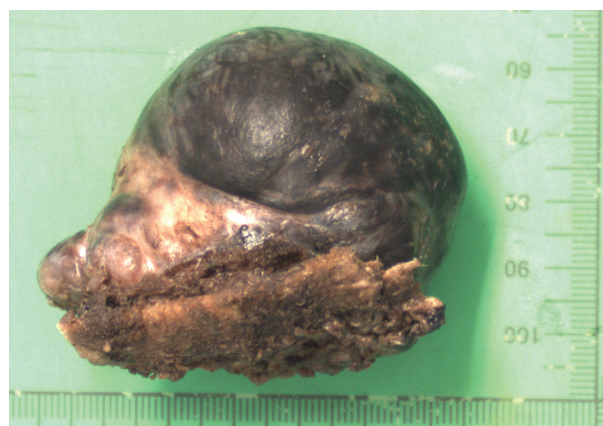


Figure 3. Surgical specimen measuring 55 × 45 × 42 mm showing the tumour with adjacent rib segment.

and hormonal changes [5]. Our patient had a history of a benign fibrocystic mass in the right breast, which was excised 6 years before her current presentation. It is unclear whether this is related to her desmoid tumour.

Wide local excision with clear margins is recommended as the main treatment option. However, it is rather challenging due to the aggressive nature of the tumour. Complete excision might also be difficult due to critical structures adjacent to the tumour. Adjuvant therapy should be considered for positive margins with close follow up in view of high rates of recurrence.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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