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Thoraco-cervical fibromatosis: treatment by surgical excision

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Abstract
We present a case of massive thoraco-cervical fibromatosis treated by sternotomy and simple excision. The patient remains disease-free after careful follow-up of three years. We suggest that if the lesion is encapsulated and position or size prevents en bloc removal, simple excision may be curative.

Key words: Fibromatosis, thoraco-cervical

Introduction

Fibromatosis ('desmoid tumour', 'desmoid fibromatosis') is a rare non-metastasizing but locally aggressive lesion arising from the musculo-aponeurotic tissue. Although well recognized in the retro-peritoneum and mediastinum it is rarely recorded in the head and neck. As this tumour has a tendency to invade local structures, removal usually involves wide en bloc surgical excision with healthy tissue at all margins. Fibromatosis arising in such a restricted anatomical region as the neck or superior mediastinum often necessitates sacrifice of neural or vascular structures to ensure adequate clearance. In some cases the tumour cannot be extirpated.

In the case described below, the tumour proved to be very large but was not invading the soft tissues of the neck or thorax apart from the posterior surface of the manubrium, where it proved to be adherent. The lesion which appeared encapsulated was removed, following sternotomy, by simple blunt dissection and excision. The area where it was adherent to the manubrium was removed in continuity with the underlying bone.

The purpose of this paper is to suggest that simple excision of this tumour may be curative in cases where the lesion fails to invade local structures and wide surgical excision is not feasible.

Case report

A 63-year-old gentleman was referred to the outpatient department with a six-month history of a gradually enlarging mass in the right side of his neck. This mass had enlarged from an initial size of 1 cm × 1 cm to a size at presentation of 12 cm × 9 cm × 7 cm. He complained of intermittent pain down his right arm which became cyanosed on exertion. His trachea was deviated left of midline but there was no airway compromise. Computed tomography (CT) scan demonstrated the mass extending from the cricoid cartilage inferiorly to the aortic arch, with postero-lateral displacement of the superior vena cava and innominate arteries (Figure 1). The patient proceeded to theatre and a combined approach was taken via standard neck incision and mid-line sternotomy. At operation the mass appeared encapsulated and free from all surrounding tissue apart from an area adhering to the posterior surface of the manubrium and sternoclavicular joint. The mass was easily removed from the neck and mediastinum following sternal splitting (Figure 2). The area of the lesion adherent

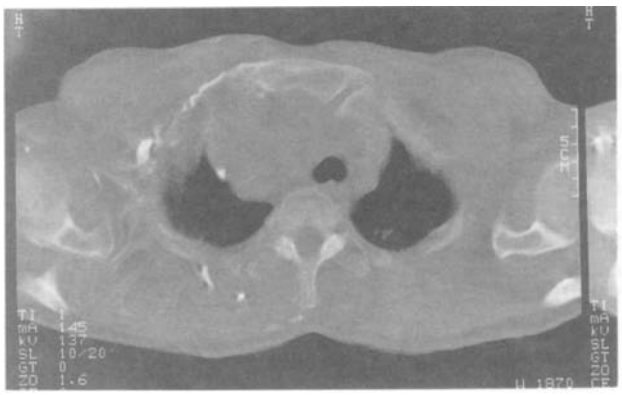

Fig. 1
CT demonstrates large mass in upper mediastinum.

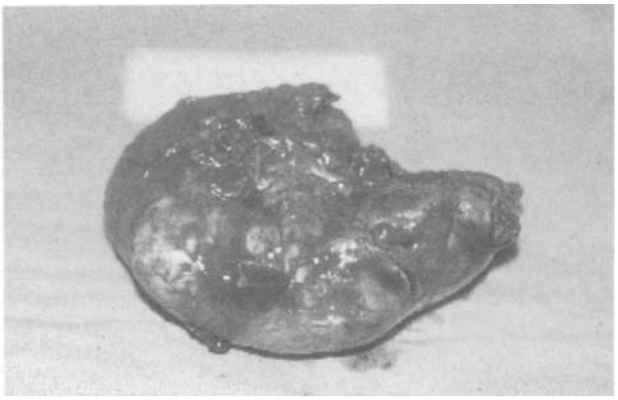

Fig. 2
Large fibromatosis (12 cm × 9 cm × 7 cm) excised from mediastinum following sternotomy.

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to the manubrium and sternoclavicular joint was removed in continuity with the underlying bone requiring resection of 2 × 3 cm sternum.

The patient made an uneventful post-operative recovery and has no recurrence demonstrable on yearly CT scan over the past three years.

Discussion

This case presented some interesting problems in the management of this rare tumour. The large size of the lesion and extension into the mediastinum required a combined thoracic-neck approach to gain access to the tumour and control of the great vessels. During surgery it was clear that the lesion was not invading the soft tissues of the mediastinum and the tumour lifted easily out of the thorax following sternotomy. The sternal split was required as the greatest dimension of the tumour was wider than the thoracic inlet.

Aggressive fibromatosis usually invades the surrounding local tissue and this invasion is seen at a microscopic level. Inadequate removal is associated with local recurrence and this may be seen in up to 70 per cent of cases (Masson and Soule, 1966). For this reason it is generally felt that complete surgical removal requires excision of the tumour en bloc using the technique of radical neck dissection (Das Gupta *et al.*, 1969; Siemssen and Anagnostaki, 1984). In this case however the tumour appeared encapsulated and free of the surrounding soft tissue structures. The size of the tumour and its close proximity to the vital structures of the mediastinum and neck clearly prevented an en bloc resection with healthy tissue at all margins. It was felt at operation that the tumour could be completely removed by simple excision. The only area of concern was the point of tumour adherence to the manubrium at the sternoclavicular joint. A margin of bone was removed in continuity with the lesion at this point.

Immunohistochemistry confirmed the diagnosis of fibromatosis and histology of the tumour, as expected, was seen to extend to the resection margin. Recurrence times of head and neck desmoid tumours are reported from two months to eleven years (Fu and Perzin, 1976; Leibel *et al.*, 1983) and although this patient has remained disease free for three years we shall need to monitor him carefully in the future.

In summary, we feel that this unusual case illustrates the point that if these lesions appear encapsulated and are of such a size that prevents en bloc removal, simple excision of the mass may be curative.

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