Recurrent large thoracic desmoid

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ABSTRACT

Desmoid tumours, also termed aggressive fibromatosis, are rare soft tissue neoplasms derived from fascial or musculo-aponeurotic structures. These tumours are histologically benign, but may behave aggressively. Problems in management are encountered because of their potential to attain large sizes, recur, and infiltrate neighbouring vital structures. Wide excision with adequate margins is the treatment of choice. Adjuvant or rarely primary radiation has a defined role. We report a 37-year-old man with a large recurrent thoracic desmoid which attained a very large size.

Keywords: aggressive fibromatosis, desmoid tumour, fibromatosis, recurrent desmoid tumour, thoracic desmoid

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INTRODUCTION

Desmoid tumours, also termed aggressive fibromatosis, are rare soft tissue neoplasms derived from fascial or musculo-aponeurotic structures. They constitute less than 0.03% of all neoplasms.[1] They are mainly found in the limb girdles and proximal extremeties, neck, trunk, abdominal wall, bowel wall and mesentery.[2] These tumours are histologically benign, but may behave aggressively at the local level, with multiple recurrences being common.[3] For extra-abdominal desmoids, local recurrence rates range from 24% to 77% in reported series.[2,4] Problems in management are encountered because of their potential to attain large sizes, recur, and infiltrate neighbouring vital structures. Reports of thoracic desmoid are very infrequent.[5] We report a case where the tumour had a repeated recurrence over an 11-year period and was extremely large.

CASE REPORT

A 37-year-old man presented with a large mass protruding through the thoracic cage and the neck (Fig. 1). 11 years earlier, the patient underwent an excision of a swelling measuring about 2 cm in size, situated near the medial end of the left clavicle. No intrathoracic extension was recognised. Excision biopsy was reported as a desmoid tumour. Three years later, patient presented with chest heaviness and was evaluated. There was a large mediastinal mass. An excision was attempted through a median sternotomy. The mass was found to be infiltrating the great vessels and the pericardium. It was excised by shaving it off the great vessels. Adjuvant radiation was planned in view of such extensive disease. The patient received postoperative radiation up to 50 Grays in 25 fractions daily for five days a week over five weeks, with telecobalt to the mediastinum with relative sparing of the cardia and lungs. Six years later, the patient had tightness in the chest but was reluctant for any medical evaluation. He presented two years later with this large mass, eight years since sternotomy (Fig. 1). At this stage, the patient was unfit for any evaluation or definitive treatment, and was confined to bed. Three months later, he succumbed to the disease.

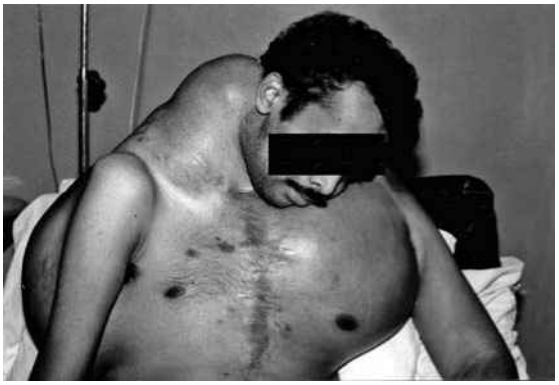

Fig. 1 Photograph shows a large recurrent intrathoracic desmoid protruding through the anterior chest wall on the left, and neck and posterior chest wall on the right. The previous sternotomy scar is also seen. The tumour measured 80 cm in its anteroposterior extent.

DISCUSSION

The term "desmoid" originates from the Greek word "desmos", meaning band or tendon. Initially, it was used to denote fibrous tumours of the abdominal wall in parous women. It was later realised that this tumour could also develop in other parts. Aggressive fibromatosis is a mesenchymal neoplasm and develops from the muscle connective tissue, fasciae and aponeuroses. It is a locally invasive but non-metastasising soft tissue lesion composed of clonal proliferation of spindle (fibrocyte-like) cells, and is characterised by a high risk of recurrence (25%–65%) after surgical treatment.[4]

Sporadic desmoid tumours have been estimated to occur in 2–5 persons per 1,000,000 population per year. It could also be associated with familial adenomatous polyposis or Gardner's syndrome. The female-to-male

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ratio of occurrence vary from 2:1 to 5:1.[4] The majority of patients with extra-abdominal desmoid are between the ages of 15 and 45 years at the time of diagnosis.[6] The proposed "DES" classification based on diameter, expansion and location is not widely accepted.[4] Intrathoracic desmoid tumours are extremely rare, with only 12 cases having been reported in the literature till 1994.[5] Patients with these lesions are often asymptomatic, and commonly present with lesions greater than 10 cm in size.[3] Although the aetiology of desmoid tumours is poorly defined, trauma, endocrine and genetic factors have been considered as causative factors.[4] Trisomies of chromosome 8 and 20 have been described, which have an increased risk of recurrence and more aggressive clinical behaviour.[7] Derangement of apoptosis gives desmoid tumour cells a proliferative advantage, and presumably, forms the basis of its high recurrence rate. Therefore, inhibitors of apoptosis proteins may be useful for predicting recurrence.[8]

It is generally accepted that wide surgical excision with the margin of clean tissues (2–3 cm), remains the principle therapeutic manoeuvre. However, this is at times impossible to achieve without sacrificing vital structures resulting in incomplete resections, leading to recurrences and poor long-term outcomes. Another contradicting view is that wide excision need not, or rather, should not be performed. High postsurgical recurrences coupled with higher aggressiveness have led to more conservative therapeutic management, stating that less radical excision is adequate with a view of optimal function. Presence of residual disease cannot be clearly shown to impact adversely on five-year disease-free or overall survival.[2] In the present case, the second surgical excision was never aimed to achieve negative surgical margins, but was aimed at preserving structure and function of the vital organs.

Some studies have shown no impact of additional radiotherapy or chemotherapy over local control of the disease.[1,9,10] Hence, they suggest re-excision for recurrence.[3] The role of radiation therapy in an adjuvant setting, if any, is limited to the control of local recurrences.[11] The role of adjuvant radiotherapy in patients with positive margins following resection of the primary disease is controversial, and should be based on potential morbidity from radiotherapy compared to the potential morbidity of another local recurrence. Some data suggests that postoperative radiation with 50–60 Gy may improve progression-free survival in incompletely-excised tumours with acceptable morbidity.[11,12] However, there is no difference in disease-specific survival.[13] It is equally important to emphasise that there are adequate large studies showing usefulness of radiation in desmoid tumours.[14-17] All these studies have shown usefulness in adjuvant postsurgical setting or primary radiation for unresectable or recurrent disease. One important conclusion is that adjuvant radiation therapy can offset the adverse impact of positive margins.[14] In addition, radiation alone can be used to good effect and durable results for unresectable or patients unfit for surgery.[15] Unfortunately, though the numbers are large, these studies have very little data on intrathoracic desmoids, reflecting the rarity of occurrence at this site. This patient received adjuvant radiation after his second surgery. In a prospective cohort study, neoadjuvant chemotherapy with doxorubicin and radiation followed by surgical resection, was found to provide excellent local control, even in those with recurrent disease.[18] Methotrexate plus vinblastine given every 7–10 days for several months is associated with prolonged stable disease in a substantial subset of patients with advanced (inoperable) aggressive fibromatosis.[19] Non-surgical methods, such as application of antioestrogens and nonsteroidal anti-inflammatory drugs, are based on limited clinical material.[4]

In conclusion, thoracic desmoid is an extremely rare entity and this is one such very rare instance to be reported. This case illustrates that long survival (11 years) can be followed by rapid death, despite the best of surgical and radiation therapies. As typical of desmoid tumours, recurrences are known. In view of the vital thoracic structures, standard three-dimensional wide excision would not be feasible. This would result in recurrences which would be more difficult to manage. This patient had a recurrence which is probably one of the largest to be compatible with life. Chemotherapy and radiation can be considered in select cases. Unfortunately, this patient developed an extremely large recurrence postsurgery and postradiation that was not amenable to any form of therapy, and succumbed to the disease. With recent availability of linear accelerator and conformal radiation, better radiation results could be expected in future.

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