CASE REPORT

Desmoid tumours: 3 cases involving the root of neck and literature review

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Abstract

Desmoid tumours are a group of rare soft tissue tumours of mesenchymal origin, which arise following the monoclonal proliferation of well-differentiated fibroblasts. They are highly infiltrative and can be locally recurrent, but do not metastasize and may stabilise with time. This article describes 3 cases of desmoid tumours in a junctional area, and a literature review in the clinical management of this entity is also presented.

Three patients with junctional desmoid tumors were identified from the institutional soft tissue database comprising of 94 cases between 1994 and 2007. The case files were reviewed and the patient histories were summarized.

All 3 were female, and all presented with arm symptoms. Resection margins were either microscopically positive or narrowly cleared at the primary surgery of all 3 patients. Two patients suffered from recurrence, but all were able to achieve long-term survival of at least 12 years.

The management of desmoid tumours is evolving with increasing knowledge of the natural history of this disease. Watchful waiting is increasingly utilized for asymptomatic or stable cases. For symptomatic or progressive disease, treatment options include surgical resection, radiotherapy and systemic therapy. Both radiotherapy and systemic therapy have been used as primary treatment modalities, neoadjuvant therapy or adjuvant therapy with varying degrees of success. Due to the complexity of this disease, and the unpredictable natural history, it is recommended that all patients with desmoid tumors be managed in a multidisciplinary setting.

Key words

Desmoid tumour, Desmoid, Aggressive fibromatosis

1 Introduction

Desmoid tumours are a group of rare soft tissue tumours of mesenchymal origin, which arise following the monoclonal proliferation of well-differentiated fibroblasts [1-3]. There are an estimated 2 to 4 cases per million people per year [4]. They represent 0.03% of all tumours and less than 3% of all soft tissue tumours [5].

Whilst classically considered to be benign, they can often be locally aggressive, invading into surrounding tissues and recurring after excision. The most important risk factor for recurrence is an involved excision margin [5]. The ability to achieve clear resection margins is heavily dependent on the anatomical location of the mass. Desmoids can be anatomically classified into three groups: intra-abdominal, extra-abdominal and abdominal wall. Of these three groups, extra-abdominal desmoids are often the most technically challenging to resect [5].

This paper describes three cases of extra-abdominal desmoid tumours occurring in a junctional region. Clear excision margins are especially difficult to obtain owing to the complexity of the anatomical location of this subgroup of tumours. A literature review is also presented to accompany the case reports.

2 Methods

A total of 94 cases of desmoid tumours were identified in the prospectively maintained, institutional soft tissue surgical database, between 1994 and 2007. Of these, three cases were junctional desmoid tumours. In this manuscript, a tumour was considered to be in the junctional region if it involved the root of the neck, at least partially occupying either the thoracic inlet or axillary entrance. Demographic and clinical data including presentation, management and follow-up were obtained from case files.

3 Results

Three (3%) of the 94 patients were found to have junctional desmoid tumours in the institutional database. These 3 cases are presented in this report.

Patient 1: A 45 year-old female machinist presented with left arm swelling and was found to have a mass on the left upper chest wall. The 2.5 cm mass was hard on palpation, fixed to the upper chest wall, and had poorly defined margins. The patient underwent a wide local excision of the upper left anterior chest wall, including partial resection of the 1st and 2nd ribs, the mid-portion of the clavicle, and the lateral 1 cm of the manubrium. The tumour was dissected free from the subclavian vein, and the defect was reconstructed with a rotational pectoralis major flap. On histology, the tumour was 45 mm × 35 mm × 22 mm in size, and composed of spindle cells with collagenous stroma, consistent with desmoid tumour. Despite close but clear margins, the patient developed a symptomatic recurrence 12 months later with extensive infiltration, involving the scalenus anterior muscle, the vertebral and the subclavian arteries, and surrounding cords of the brachial plexus. The recurrent tumour was deemed unresectable without a fore-quarter amputation, and was therefore managed with radiotherapy (63 Gy in 35 fractions). A favourable response was seen with the radiotherapy, and on follow-up imaging, there was no evidence of residual disease. The patient is currently disease-free at 19 years following the radiotherapy for recurrence, with no significant late radiation toxicity.

Patient 2: A 62 year-old housewife presented with neck and right arm pain, and axial imaging revealed a 5 cm tumour overlying the lower right trapezius muscle, in the posterior triangle of the neck. It was hard on palpation, but appeared mobile. The patient was initially treated at a regional centre, undergoing a wide local excision, including partial resection of the right trapezius and levator scapulae muscles. The histology showed a 53 mm × 27 mm × 26 mm spindle cell tumour, consistent with desmoid tumour, with microscopic positive margins. At 20 months from her initial surgery, the patient re-presented with another mass at the same location to the authors' institution. This was treated with re-excision of the mass, followed by adjuvant radiotherapy (50 Gy in 25 fractions). The histology confirmed recurrence of a spindle cell tumour of 90 mm × 35 mm × 30 mm in size, with extensive skeletal muscle infiltration, and microscopically positive margins. At 11 months after her 2nd resection, she again re-presented with local recurrence in the posterior triangle on the right side of her neck. This core biopsy confirmed recurrent desmoid tumour was deemed unresectable as it was clinically fixed, deep to the upper 3rd of the sternocleidomastoid muscle. The patient was treated with tamoxifen (20 mg daily) for

15 months – ceased after developing deep vein thrombosis. At the time of report, the patient has been followed for 12 years, and was alive with clinically stable disease.

Patient 3: A 22 year-old female clerk presented with persistent left arm swelling following a fractured humerus from a motor vehicle accident. Axial imaging revealed a tumour in the left supraclavicular fossa. The tumour was well-circumscribed, 4 cm in maximal dimension, and clinically fixed to the clavicle. An incisional biopsy confirmed a desmoid tumour, and a wide local excision was subsequently performed, including partial removal of the pectoralis major muscle and the medial two-thirds of the clavicle. The histology again confirmed a spindle cell tumour of 35 mm in maximal dimensions, consistent with desmoid tumour, with microscopically positive margins. She was also treated with adjuvant radiotherapy (63 Gy in 35 fractions), and tamoxifen (20 mg daily) for 2 years. Currently, the patient is disease free at 18 years of follow-up, with no significant late radiation toxicity.

4 Discussion

The three cases presented in this article have many common features. All three patients were female, and they all presented with symptoms affecting the arm due to a desmoid tumour in the junctional region of the neck, arm, and thorax. Due to the anatomical location, resection margins were either microscopically positive or narrowly cleared at the primary resection of all 3 patients. Two of the patients suffered from recurrence, but were able to regain long-term disease control (12 to 19 years). The age at first presentation ranged from 22 to 62, and recurrence was variably treated with surgery, radiotherapy, and/or tamoxifen.

4.1 Epidemiology & Genetics

Desmoid tumours are rare. Its reported incidence ranges from 2 to 4 cases per million people per year. Desmoid tumours can occur at any age, but are most commonly diagnosed between the ages of 10 and 40 years. They are slightly more common in women than men, and occasionally occur in association with pregnancy or trauma. Most desmoid tumours arise sporadically; however, a minority are associated with mutations of the familial adenomatous polyposis (FAP) or adenomatous polyposis coli (APC) genes. Approximately 85% of the spontaneous desmoid tumours are associated with beta-catenin mutations, which cause defects of the Wnt/catenin signaling pathway [6-11].

4.2 Pathology

Desmoid tumours are a heterogeneous group of monoclonal, myofibroblastic neoplasms with a propensity for local recurrence, but not metastases. Histologically, the cells appear stellate to spindle-shaped within a collagenous matrix. Without a capsule, these cells often demonstrate an infiltrative growth pattern into the surrounding tissue, typically soft tissue and skeletal muscle. However, they differ from classic malignant cells as they lack the pleomorphic, atypical, or hyperchromatic nuclei [12]. Also unlike classic malignant tumours, they do not metastasize, despite often being locally invasive and/or multifocal.

4.3 Presentation

Desmoid tumours can arise from any musculoaponeurotic sheath in any part of the body, and are commonly classified by anatomical locations as intra-abdominal, extra-abdominal or abdominal wall tumours. Junctional desmoid tumours are a subset of extra-abdominal desmoid tumours. This subset of desmoid tumours warrants specific discussion due to the associated difficulty in obtaining clear resection margins and hence additional challenges in management. Pain, mass, and swelling of the associated limb are the commonest symptoms on presentation.

4.4 Management

As with all malignant or rare diseases, patients with desmoid tumours are best managed in a multidisciplinary setting within a referral centre. In this article, the discussion on management focuses on extra-abdominal desmoids, with

emphasis on junctional tumours where evidence is available. Unlike abdominal wall desmoid tumours, which are associated with long-term disease-free survivals of over 90%, extra-abdominal desmoid tumours are associated with a poorer prognosis – less than 40% of patients achieving long-term disease-free status [6].

In general, the clinical course of desmoid tumours can be unpredictable, and different outcomes have also been reported between tumours of differing anatomical locations [5]. Some tumours are rapidly destructive; while others remain dormant for many years without treatment; and yet others naturally regress [13-15]. This unpredictability in its natural course has created challenges in establishing management guidelines. Historically, aggressive management has been adopted involving a combination of radical surgery and radiotherapy. More recently, several series have reported that up to 65% of asymptomatic patients with desmoid tumours did not show progressive disease over 3 to 6 months of observation, validating "watchful waiting" as an accepted management option [15].

In 2013, Fiore et al reported 97 cases of pregnancy related desmoid tumours. Of the patients initially managed with watchful waiting (43%), 37% required no further treatment and 14% had spontaneous regression [16]. In addition, Eastley et al published a retrospective review of 47 desmoid tumours, inconsistently managed with respect to imaging, biopsy technique, involvement of multidisciplinary teams and treatment undertaken. Their overall recurrence was 19% recurrence after 4.9 years of follow-up [17]. Both of these papers demonstrate that disease free survival is not necessarily related to complete surgical resection as previously understood.

In patients with progressive disease, treatment usually consists of surgery, external beam radiotherapy, and/or systemic therapy, in various combinations. However, the success of selecting the appropriate modalities depends on careful assessment of the type and extent of disease.

4.5 Initial assessment

Initial assessment of a tumour that is suspected to be a desmoid tumour should include a search for clinical features of Gardner's Syndrome (triad of colonic polyposis, osteoma, and soft tissue tumours) in addition to general history taking, and physical examination to determine the clinical features of the mass, as well as the secondary effects. Following clinical assessment, appropriate axial imaging (CT, MRI, or both) of the involved body region is invaluable in correlating to the clinical findings, and to help determine resectability. While magnetic resonance imaging (MRI) provides good soft tissue details, computed tomography (CT) may also be required to define vascular and bony involvement [18].

Following clinical and imaging assessments, a carefully planned, image guided core or incisional biopsy are often necessary to confirm the diagnosis of desmoid tumour before further management decisions can be made. When planning the biopsy approach, it is important to consider the ultimate surgical approach, in order to facilitate incorporation of the biopsy track into the surgical incision. In select circumstances, histological confirmation may be omitted if resection with a clear margin can be easily achieved.

4.6 Surgery

There is no doubt that the aim of surgery is to achieve tumour debulking. However, microscopically positive resection margins may be acceptable if excessive morbidity would result as a consequence of achieving negative margins. Striking a balance between surgical clearance and morbidity is especially pertinent in extra-abdominal tumours, where definitively clear margins may require the sacrifice of significant limb function. Whilst desmoid tumours recurrences have been reported despite complete excision, long-term survival can be achieved in the context of positive resection margins, as illustrated by the cases in this article [19]. However, other investigators have suggested that surgical margin is a statistically significant factor affecting local recurrence [20].

4.7 Radiotherapy

Radiotherapy has a role in both the management of unresectable or inoperable and resected disease [21]. In patients who have disease, either primary or recurrent, that is unresectable without unacceptable morbidity, radiation alone appears to provide very adequate local control of up to 78% in one review [22]. In patients who have had surgery, radiotherapy (at a dose greater than 50Gy, with a fraction size of ≥ 2Gy) significantly improved local control compared to patients treated with surgery alone, and adjuvant radiotherapy appears to be better than treating with radiotherapy when patients recur later [20, 23]. The influence of positive margins after surgery appears to vary in different studies but it does appear that radiotherapy improves local control in patients with positive margins to levels similar to those in patients with clear microscopic margins [20, 22]. Complications after radiotherapy are dose related.

4.8 Systemic therapy

Systemic therapy is generally reserved for patients who are not amenable to surgery or radiotherapy, or those with locoregional recurrences. The modalities include endocrine therapy, non-steroidal anti-inflammatory drugs (NSAIDs), tyrosine kinase inhibitors, and some conventional chemotherapy. Only a brief discussion is given here [24]. Comparative studies are lacking and at present, most data regarding the use of systemic therapy comes from case reports.

Endocrine therapy is based on the apparent sensitivity of desmoid tumours to sex hormones, especially the oestrogens. Growth is often observed in pregnancy or after menarche [25], with regression at menopause [26-29]. Extra-abdominal desmoid tumours have also been observed to fluctuate in size with the menstrual cycle. These observations are thought to result from the presence of oestrogen receptors (ER). Most investigators have shown that desmoid tumours are positive for ER-β receptors, but negative for ER-α [30, 31]. Tamoxifen is the most common endocrine agent used in the treatment of desmoid tumours. It is a selective ER modulator that inhibits cell proliferation *via* ER binding [32]. Tamoxifen can induce some response in about 50% of treated patients [33]. It is prudent to confirm the ER status of a tumour prior to initiating endocrine treatment. Dosing is the same as secondary prevention for breast cancer, although there is some anecdotal evidence for efficacy at higher doses [33, 34].

Aside from tamoxifen, a few other agents have been used to stablise desmoid tumour progression with varying success, including non-steroidal anti-inflammatory drugs (NSAIDS), interferon, and selective tyrosine kinase inhibitors such as imatinib. Traditional cytotoxic chemotherapy is reserved for patients with rapidly progressive symptomatic disease, after failing hormonal therapy. Several regimes are utilised. Limited success has been reported using single agents such as the anthracyclines [35, 36].

4.9 Management of recurrence

Unfortunately, local recurrence of desmoid tumours is common, and rates between 50% and 90% have been reported [4, 25, 37]. Local recurrence tends to occur within two years of primary treatment [38]. In a large, prospective multivariate analysis, Crago et al (2013) identified tumour site, size and patient age as being independent predictors of recurrence [39]. The most likely extra-abdominal sites for recurrent desmoid tumours are: popliteal fossa, supraclavicular fossa and buttock [1, 29, 40]. Higaki et al (1995) found that whilst size was not a risk factor for recurrence, the number of muscles involved by the tumour was a reflection of its aggressiveness and likelihood to recur [40]. More recently, specific genetic mutations have been linked with primary treatment failure [11].

Similar principles apply to the management of recurrence, as for primary tumours. The histological confirmation of recurrence is usually required by a core biopsy. Progressive disease should be considered for repeat debulking surgery, if the tumour is resectable and the resection does not cause undue functional or cosmetic disability. Treatment with radiotherapy and/or systemic agents should be considered as adjuncts or alternatives to surgery. In these complex patients, management in the setting of a multidisciplinary team is advisable, and participation in clinical trials is encouraged.

5 Conclusion

The management of desmoid tumours is evolving. Whilst strong evidence in its management is lacking, the accumulated experience in case reports and case series have provided traction for watchful waiting as a valid strategy in stable disease. For progressive disease, surgical debulking with or without adjuvant radiotherapy/systemic therapy remains the recommended treatment strategy. The unpredictable nature of the disease challenges the traditional view about microscopic positive resection margins, and dictates the need for long-term follow-up of all patients. The subgroup of junctional desmoid tumours brings further challenges for clinicians owing to their anatomical location.

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