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Review

The management of soft tissue tumours of the abdominal wall

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Abstract

Background: Soft tissue tumours of the abdominal wall account for approximately 10% of all soft tissue tumours. Tumours at this site comprise a heterogeneous group of pathologies with distinct clinical behaviours and responses to treatment. The management of these tumours has largely been extrapolated from studies of soft tissue tumours at other sites. This review aims to summarise the existing data relating to abdominal wall tumours and suggest principles for managing soft tissue tumours at this site.
Methods: Relevant articles were retrieved from a comprehensive literature search using the PubMed database. Key words included abdominal wall, soft tissue tumours, surgery, radiotherapy and chemotherapy. No restrictions on publication date were used.
Results: The most common pathologies presenting in the abdominal wall are desmoid tumours, soft-tissue sarcoma and dermatofibrosarcoma protuberans (DFSP). Desmoid tumours should be managed with an initial period of observation, with surgery reserved for progressive lesions. Surgery should be the primary treatment for soft-tissue sarcomas and DFSP, with radiotherapy reserved for large-high grade tumours and preferentially given pre-operatively.
Conclusions: Abdominal wall tumours are rare and should be managed in centres with experience in the management of soft tissue tumours. Management should be tailored to the biological behaviour of specific pathologies.

Keywords: Abdominal wall; Desmoid tumours; Soft tissue sarcoma; Dermatofibrosarcoma protuberans

Introduction

Soft tissue tumours arising in the abdominal wall comprise a range of different pathologies, with distinct clinical behaviours and patterns of relapse. Tumours of the abdominal wall are rare, accounting for less than 10% of all soft tissue tumours.[1] As soft tissue tumours most commonly affect the extremities, the principles of management of abdominal wall tumours are in part extrapolated from randomised trials in extremity sarcoma, coupled with cohort studies that include abdominal wall tumours alongside soft tissue tumours at other anatomical sites. There are a relatively small number of retrospective studies of abdominal tumours studied in isolation. This review article will summarise the data that exists relating to abdominal wall tumours and suggest management principles for patients with soft tissue tumours arising at this site.

Epidemiology

The most commonly occurring tumours of the abdominal wall are desmoid tumours and soft tissue sarcomas, accounting for 45% and 40% of all abdominal wall tumours respectively, with dermatofibrosarcoma protuberans (DFSP) accounting for the remaining cases.[2–4]

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Desmoid tumours, also known as aggressive fibromatosis, are monoclonal fibroblastic proliferations that are characterised by an unpredictable potential for progression or spontaneous regression. The estimated incidence of desmoid tumours is 5/1,000,000 per year, with evidence that their incidence has increased over the past two decades.[5] Desmoid tumours lack the ability to metastasise but local progression may cause significant morbidity or even death, particularly when located within the abdominal cavity. Desmoids demonstrate an infiltrative growth pattern and are associated with high rates of local recurrence. These tumours typically occur in younger patients, with a peak incidence in the fourth decade.[5–7] Sporadic mutations in the β-catenin gene, *CTNNB1*, account for the majority of desmoid tumours, with approximately 10% of cases arising in association with an *APC* mutation in the context of the familial adenomatous polyposis (FAP) syndrome.[8–11] Although desmoid tumours may occur virtually anywhere within the body, the extremities and the abdominal wall are the most commonly affected sites.[12] Desmoid tumours occur more commonly in females, who account for over two-thirds of desmoids at any site.[5,7,12] Abdominal wall desmoid tumours show a particular predilection for women of childbearing age, who account for over 90% of patients presenting with desmoids at this site.[13,14] Current hypotheses for the increased incidence of abdominal wall desmoids in these patients include physical trauma secondary to pregnancy or, as these tumours may often express oestrogen or progesterone receptors, a hormonal component to their aetiology.[15,16] Intra-abdominal disease is most commonly seen with FAP-associated desmoids and it is important to exclude this syndrome in such patients, as the management of intra-abdominal desmoids is very different between patients with FAP and those without.[12,17,18] Surgical management of intra-abdominal desmoids in the context of FAP is associated with high peri-operative morbidity and mortality as well as high rates of local recurrence.[17,19] In contrast, surgery for sporadic intra-abdominal desmoids is associated with fewer complication or recurrences.[18] As such, in FAP-associated intra-abdominal desmoids, there is a high threshold for surgical intervention, which is generally reserved for patients who have failed medical management.

Soft tissue sarcomas represent a diverse group of cancers of mesenchymal origin with an estimated incidence of 4–5/100,000 per year in Europe.[20] The propensity for distant metastasis varies between distinct histological subtypes, although all subtypes have the potential for local recurrence.[21] Soft tissue sarcomas can arise at almost any anatomical site but most commonly present in the extremities or the abdominal cavity/retroperitoneum. Primary abdominal wall sarcomas account for less than 5% of cases.[1,3] The most common subtypes manifesting in the abdominal wall are undifferentiated pleomorphic sarcomas (previously known as malignant fibrous histiocytomas), fibrosarcomas and synovial sarcomas.[2–4] In contrast to desmoid tumours, soft tissue sarcomas do not display a preference for gender and occur later in life, with a median age at presentation of approximately 50 years.[3,22]

DFSP is a locally aggressive tumour with low metastatic potential.[23–25] Rather than representing a true abdominal wall tumour, DFSP originates from the cutaneous tissues and is typically limited to superficial structures. DFSPs have an infiltrative growth pattern, particularly in the cutaneous plane, and a propensity for local recurrence. Progression to a fibrosarcoma may occur in around 15% of patients with DFSPs, conferring a more aggressive biological behaviour, with a greater risk of involvement of deeper structures and increased metastatic potential.[25] Almost all DFSPs contain a fusion gene, *COL1A1-PDFGB*, leading to activation of the platelet-derived growth factor β (PDGFβ) signalling pathway.[26,27] As such, tyrosine kinase inhibitors targeting the PDGFβ pathway, such as imatinib, may be of therapeutic benefit in both localised and disseminated disease.

Diagnosis, staging and imaging

The principles of investigation for abdominal wall tumours are broadly similar to those of soft tissue sarcomas of other anatomical sites.[28] Due to the rarity of these lesions, patients with suspected soft tissue tumours of the abdominal wall should be referred to specialist centres benefitting from multidisciplinary teams experienced in the management of soft tissue tumours.

Patients with desmoid tumours and soft-tissue sarcomas may have similar clinical presentations. The majority of patients with either pathology will present with a mass that is typically painless. Size is a poor discriminator between these pathologies, although soft-tissue sarcomas may present with slightly larger dimensions with a median size of 6–15 cm compared with 6–8 cm for desmoid tumours.[3,4] Fixation to underlying structures is suggestive of a soft-tissue sarcoma, whereas a lesion occurring in a young female, particularly with a recent history of pregnancy, is more suggestive of a desmoid tumour. In contrast to the other pathologies, DFSPs tend to present as an indurated superficial lesion with characteristic purple or blue discolouration.

In the presence of a small (<5 cm) and superficial lesion, ultrasound may be used to identify clearly benign lesions. Larger or deep lesions or those with equivocal or suspicious ultrasound features should be further assessed with cross-sectional imaging. Magnetic resonance imaging (MRI) is probably the modality of choice for imaging abdominal wall tumours, as the improved soft tissue contrast provides better delineation of tumour margins than computed tomography (CT).[29] However, CT is still a good investigation for surgical planning for soft tissue lesions of the trunk or abdominal wall, with the advantage that staging of the lungs and assessment of intra-abdominal viscera can be performed at the same time as imaging the primary tumour, and is less prone to movement artefact that MRI. MRI is reasonably accurate in differentiating benign and malignant lesions (though not accurate enough to circumvent the need for a diagnostic core

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