Extra-abdominal desmoid tumour presented as neck swelling in the trapezius muscle

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Abstract
A desmoid tumour is a type of benign fibromatosis and can be classified into intra- and extra-abdominal tumours. Extra-abdominal tumours commonly develop in the shoulder, pelvis and chest wall. This tumour can be found in all age groups, but it is more prevalent in young female adults. There are no specific symptoms related to the tumour and it is mostly painless and shows swelling at the early stage. As the tumour progresses, variable symptoms such as pain, bowel obstruction, urinary obstruction and limitation of range of joint movement develop depending on the location of the tumour. In this case report, along with a literature review, we have described a rare case of a desmoid tumour developed in the trapezius muscle accompanying neck swelling.

Introduction
A desmoid tumour is a type of benign fibromatosis and can be classified into two subtypes, intra- and extra-abdominal, and it is mainly developed in the abdomen. This report aims to describe a rare case of a desmoid tumour developed in the trapezius muscle accompanying neck swelling.

Case report
A 23-year-old female patient visited the outpatient department with a chief complaint of left posterior neck swelling that started to develop about seven months prior to visit (Figure 1).

Computed tomography (CT) images showed a round soft tissue tumour in the left trapezius muscle, with a comparatively homogeneous density of the mass and isodensity of the trapezius muscle (Figure 2).

The mass including the trapezius muscle was removed with a 2 cm safety margin and it was $4 \times 3 \times 3$ cm in size. The resection surface was light grey or yellow in colour, and it was a relatively homogeneous solid mass containing a densely trabeculated area (Figure 3). Histopathological findings showed bundles of fibroblasts and large amounts of collagen indicating an extra-abdominal desmoid tumour (Figure 4).

The patient is on an 18-month follow-up study after the operation, and there are no signs of recurrence or metastasis.

Discussion
A desmoid tumour was first described in 1832 by John MacFarlane as arising from the anterior abdominal wall and it is a rare disease, with a reported prevalence of 2–4 in 1,000,000 per year. The tumour may be located in the abdominal wall, extremities and abdominal mesenteric. Fifty percent of the tumours are found in the abdominal wall and trunk muscle layers, 40% develop in the extremities and 10% are located in the abdominal mesenteric. The most common locations apart from the abdomen are the shoulder and pelvis.

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Histological findings of the mass. (a) Histologic analysis of haematoxylin and eosin stain (×100) demonstrated high cellularity of a well-differentiated spindle-shaped fibroblast with many collagen fibres. (b) Haematoxylin and eosin stain (×400) showed several typical mitoses.

A desmoid tumour is a mass with a hard rubbery light pink surface and unclear borderlines. It infiltrates as it progresses and there are no characteristic symptoms that are directly related to the tumour. It is usually painless and swelling can be observed at the early stage of the disease, and other symptoms can develop depending on the location and the extension of invasion into surrounding areas of the tumour. As mentioned, the diagnosis can be challenging due to the lack of symptoms in the early stages.

The genesis of a desmoid tumour is as yet unknown, but endocrinologic hormonal aetiology, previous trauma and gene mutation are believed to be the causative factors. Supporting these facts, there are published research results indicating that the prevalence is high in women of childbearing age and the growth of the tumour is accelerated during pregnancy and dystrophic changes of the tumour have been observed at menopause. It also often develops in the areas of previous surgery or injury sites 30%–80% of the tumour reportedly develops in the areas of previously operated sites. Recently, it has also been reported that the onset of the tumour is related to adenomatous polyposis coli and β-catenin gene mutations.

The diagnosis can be challenging because the tumour itself does not present any characteristic symptoms but early swelling is the main characteristic of the disease and a desmoid tumour should be clinically suspected if there are any related causative factors.

Histologically, this tumour contains identical shapes of well-differentiated fibroblasts, collagen, fusiform nucleus and light chromatin structures. The micronucleus is not clear and mitosis can be observed, but indeterminate mitosis cannot be seen. When these histological findings are observed, it must be differentiated from low-grade fibrosarcoma. It is very important to differentiate the disease from low-grade fibrosarcoma as the borderline of a desmoid tumour is unclear and infiltrates into the surrounding tissues with high recurrence rates. The uniform growth pattern, maturity of tumour cell and low-grade of mitosis are the histological differentiating points. Low-grade fibrosarcoma, inflammatory pseudotumor, other fibromatoses and scar hypertrophy should be considered as a differential diagnosis.

A desmoid tumour can easily infiltrate into the surrounding tissues and shows a high recurrence rate of 25%–65%, but metastasis is rare.

Radical total resection with a >2 cm safety margin is recommended to treat the disease, and CT and magnetic resonance image (MRI) before the surgery provides vital information to understand the extension of the tumour and its relationship with the surrounding anatomical structures. Reoperation is recommended in the case of recurrence or incomplete resection and drug therapy or radiotherapy is further recommended if radical excision or reoperation is not possible.

Tamoxifen, antiprostaglandin (sulindac), goserelin acetate, interferon, doxorubicin, dacarbazine, actinomycin D, vincristine and c-adenosine 3′/5′ cyclic monophosphate (c-AMP) modulators (ascorbic acid, theophylline, testolactone) are used in drug therapy. Radiotherapy can be selectively used in the case of recurrence or tumour invasion to nerves such as the brachialplexus.

Case report

In this case report, along with a literature review a rare case of a desmoid tumour developed in the trapezius muscle accompanying neck swelling was described.

Abbreviations list

- c-AMP, c-adenosine 3′/5′ cyclic monophosphate; CT, computed tomography; MRI, magnetic resonance image.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images.

References


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Case report


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