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Case reports

Desmoid tumours of the head and neck: A case report

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ABSTRACT

Introduction: Desmoid tumours of the head and neck, also known as fibromatosis, are rare, locally invasive benign tumours with high recurrence rate, causing considerable morbidity. Complete surgical excision of desmoid tumours is considered to be the only effective treatment.
Case report: We present a case of fibromatosis of the right posterolateral region of the neck in a 56-year-old woman who presented with right neck mass. The patient underwent complete excision of the tumour with no adjuvant therapy. No recurrence or neurological deficit was observed 2 years after surgery.
Conclusion: Although desmoid tumour is a benign neoplasm with no metastatic potential, treatment is challenging due to its aggressive, infiltrative behaviour with a tendency to recur.

1. Introduction

Desmoid tumours are a group of rare tumours arising from connective tissue of muscle, fascia or aponeurosis [1]. Although they are considered to be benign lesions, desmoid tumours are locally infiltrative and can cause extensive morbidity due to destruction of adjacent vital structures and organs [2]. Only 12 to 15% of all desmoid tumours arise in the head and neck [3,4]. The optimal treatment strategy for these patients remains unclear.

We report a case of desmoid tumour of the head and neck diagnosed and treated in our department.

2. Case Report

A 56-year-old woman presented with a history of right neck mass for 9 months that was gradually increasing in size. On examination, she presented hard swelling, fixed-to-the-skin and deep neck structures, in the right posterolateral region of the neck, extending from the retroauricular region to the suprasternal notch.

Contrast-enhanced computed tomography (CT) was performed (Fig. 1) and biopsy of the mass demonstrated fibromatosis.

On the basis of radiological and histopathological findings, wide surgical excision of the mass was performed, including all the sternocleidomastoid muscles, while preserving the spinal accessory nerve. Histopathology revealed a proliferation of uniform fibroblasts in a collagenous stroma, with infiltration of adjacent adipose tissue and skeletal muscle.

No local recurrence was observed on physical examination and CT after a 2 years' follow-up (Fig. 2).

3. Discussion

Desmoid tumours are benign fibrous tumours that arise from musculoaponeurotic structures [3]. They are locally invasive, with a high recurrence rate after resection, but with no malignant potential [3,4].

Desmoid tumours commonly have a female predominance. They can occur at any age with extreme ages of 15 to 60 years in the literature [1].

The head and neck are the commonest sites of desmoid tumours, mostly (80%) involving the neck followed by the face, oral cavity, scalp, and paranasal sinuses [5].

Unlike other benign tumours of the head and neck, desmoid tumours are known to be locally invasive. This particularity is due to the dense anatomy of the head and neck region and the close anatomical relations with neurovascular and other vital structures (trachea, brachial plexus, etc.) [2,5]. Invasion of these structures may be associated with functional sequelae or even mortality [3].

The wide surgical excision of the lesion is the treatment of reference. However, the infiltrative character of the tumour in the head and neck region may cause difficulties for complete surgical resection which can be associated with an excessive risk of mortality or functional sequelae [3]. For these reasons, surgery may need to be followed by postoperative radiotherapy to control residual or recurrent disease, despite the benign nature of desmoid tumours [3].

Nuyttens et al. (2000)[6] conducted a comparative review of 22 articles on the treatment of desmoid tumours from 1983 to

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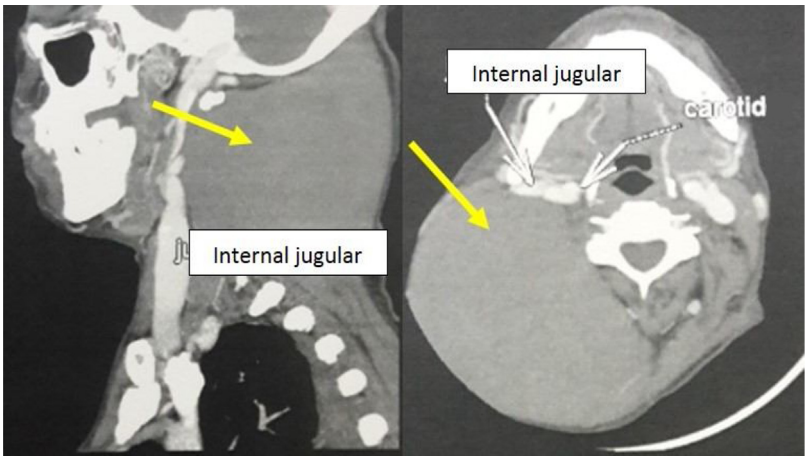

Fig. 1. Computed tomography scan of the neck showing a soft tissue mass in the upper neck lateral to the sternocleidomastoid muscle, infiltrating the deep lateral neck muscles and surrounding superficial and deep fat.

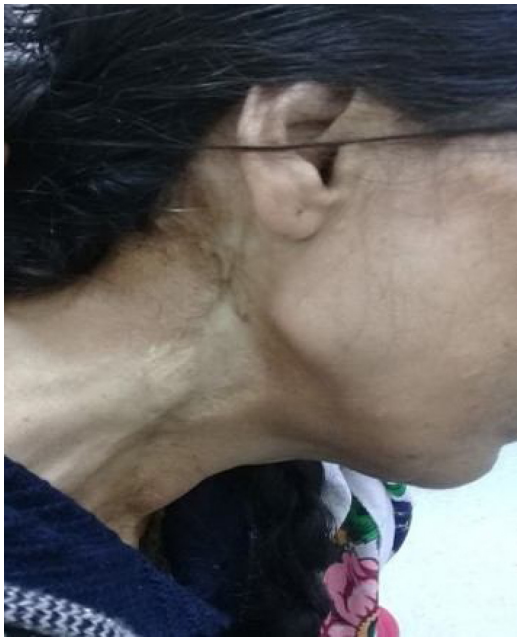

Fig. 2. Photo of the patient demonstrating absence of local recurrence 2 years after surgery.

1998 and noted that local control rates after surgery and adjuvant radiotherapy in the group of tumours with positive margins were 75%, significantly better than for the group of tumours with positive margins treated by surgery alone (41%). However, these results remain controversial and other studies have failed to demonstrate the efficacy of adjuvant radiotherapy for tumours following incomplete surgical resection [7,8].

Chemotherapy can be considered for unresectable tumours, or for patients who are unable to support the morbidity of surgery and radiotherapy [7]. Sze et al. (2009)[8] reported significant tumour reduction with low-dose methotrexate and vinblastine.

Several other agents have also been used in the treatment of desmoid tumours, such as hormonal therapy and non-steroidal anti-inflammatory drugs (NSAID) [9,10]. Meloxicam, a COX-2 inhibitor NSAID, has been shown to be effective in controlling neck and other extra-abdominal desmoid tumours [9]. The overall response rate to NSAID in combination with other medical therapy (such as antiestrogen therapy: tamoxifen) reported in the literature is about 50%, and NSAID is usually considered to be the first-line medical treatment because of its low toxicity [10]. The use of various antiestrogen agents may induce regression of desmoid tumours, as tumour proliferation, especially in intra-abdominal sites, has been observed in response to estrogens [10].

Other drugs are currently under development, such as imatinib with a response rate of less than 10% for solid tumours [10].

In some cases, a simple wait-and-see approach has been proposed for tumours not associated with any functional sequelae or with no significant risk of mortality. This approach is supported by a number of cases of spontaneous regression [3].

Patients require close follow-up due to the unknown natural behaviour of these tumours. The final decision whether to treat or not to treat should be based on the patient's symptoms, medical status, and the biological properties of the tumour [10].

4. Conclusion

Although desmoid tumour of the head and neck is a histologically benign neoplasm, this site represents a therapeutic challenge because of the infiltrative nature and the proximity of vital structures. The most effective treatment is complete surgical excision with negative surgical margins. Alternative treatment modalities, such as primary radiotherapy and medical treatment or a wait-and-see approach, are all reasonable options in selected cases and may be preferable to mutilating surgery.

Disclosure of interest

The authors declare that they have no competing interest.

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