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Case Report

Nodular fasciitis affecting the masseter – A rare and rapidly growing mass

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ABSTRACT

Nodular fasciitis is a rare condition characterised by rapid growth of fibroblasts and myofibroblasts. Most reported cases occur in the torso and upper limbs, with head and neck nodular fasciitis being an exceedingly rare entity.

Due to its locally aggressive and rapidly growing nature, nodular fasciitis can be mistaken both on clinical and histological grounds as a malignant process. Despite this, definitive management is via surgical excision, and recurrence is unlikely.

A case of facial nodular fasciitis is described, with reference to the current understanding on its clinical presentation, radiological and histological features, and management.

Nodular fasciitis is an important differential when considering fast growing facial masses. Misdiagnosis as malignancy on clinical or histological grounds can lead to unnecessary wide excision, which is disfiguring and grossly unnecessary for this benign condition.

1. Introduction

Nodular fasciitis (NF) is a rare and rapidly growing tumour affecting the muscular fascial layers. It is thought to represent a benign process that is likely to be reactive to irritation or trauma [1]. Due to its rapid growth and mimicry of malignancy, NF has also been termed pseudosarcomatous fasciitis, infiltrative fasciitis, or pseudosarcomatous fibromatosis [1].

Nodular fasciitis most commonly affects the upper limbs and torso [1,2]. Some cases of head and neck NF have been reported, compromising of between 7–20% of all NF cases [3]. Of all head and neck NF, the majority occur in the oral cavity [3]. When affecting the face, NF is usually described over the bony prominences of the mandibular angle or zygoma [1]. NF has no gender predilection, and tends to occur in the third to fifth decades of life, with an average age on diagnosis of 35 years [3]. The exception to this is in maxillofacial NF, which is described to occur more commonly in children [3].

Nodular fasciitis of the masseter in an adult is a rare entity. Due to its rapid growth and histological features, it can mimic aggressive malignancies such as head and neck sarcoma. For this reason, an understanding of nodular fasciitis affecting the masseter is crucial, such that inadvertent resection is avoided.

2. Case report

A forty-seven-year-old female presented with a one-month history of a left masseteric mass. This mass was reported to be minimally symptomatic, and was non-tender unless she was chewing. When first noticed one month prior to her presentation, the lesion was reported to be soft, mobile, and did not restrict her jaw movements whatsoever. She had no relevant medical history and there was no history of preceding trauma to the area.

The lesion presented as a well localised lump that was palpable bimanually and was more obvious on mouth opening. The mass measured approximately 20 × 20 mm in dimension and appeared to be intramuscular (Fig. 1). There were no overlying skin changes, the swelling was firm and was not tethered to overlying skin or underlying mucosa.

An ultrasound guided fine needle aspiration was performed, which revealed spindle and epithelioid cells, but was inconclusive.

An MRI was performed (Fig. 2) showing a solitary lesion within the left masseter. The lesion was reported as representing a benign process such as a Schwannoma, but a more aggressive process was also suggested. The mass was isointense to muscle on T1 imaging, and hyperintense on T2. There was no cortical erosion of the adjacent mandible, and a small area of cystic degeneration could be seen centrally. Some restricted diffusion was seen around the lesion. Due to the benign

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Fig. 1. Clinical photographs. (a) Photograph showing well localised left cheek lump. (b) The lump is more apparent on mouth opening.

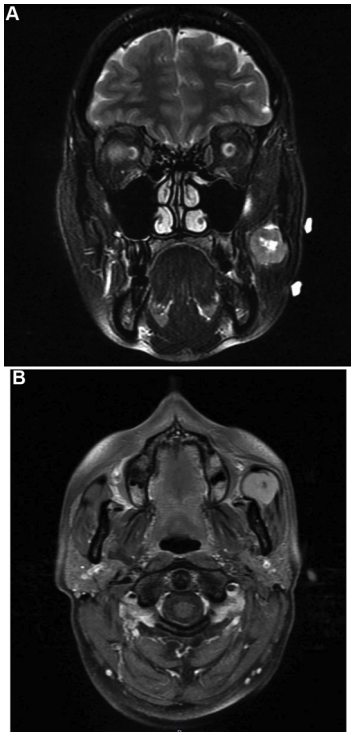

Fig. 2. MRI of left masseteric lesion. (a) T2 fat saturated coronal view showing hyperintense signal from left masseteric mass with evidence of central cystic degeneration. (b) T1 fat saturated axial view showing isointensity to muscle and central cystic degeneration.

appearance on MRI, a decision was made to perform an excisional biopsy of this lesion.

The patient underwent an excision of the lesion via a trans-oral approach, under general anaesthetic. The excision was performed in an extracapsular plane, maintaining a thin cuff of muscle around the lesion to preserve integrity of the mass and ensure complete removal (Fig. 3). Histopathology confirmed nodular fasciitis, seen as a solid nodular spindle cell lesion. There were spindled fibroblastic cells arranged in a herringbone pattern. Areas of hypercellularity were interspersed with areas of myxoid stroma, extravasated erythrocytes and scattered lymphocytes (Fig. 4). Immunohistochemistry showed positive staining of

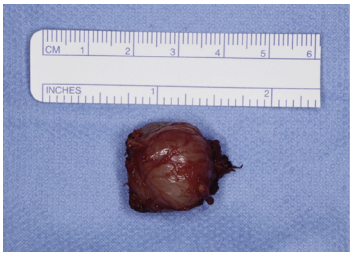

Fig. 3. Excised specimen – well circumscribed solid lesion approximately 20 mm in maximum diameter.

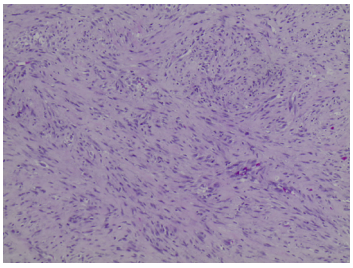

Fig. 4. Haematoxylin and eosin stain demonstrating loosely arranged bundles of spindled fibroblastic/myofibroblastic cells without significant atypia, and scattered small-sized thin-walled vessels.

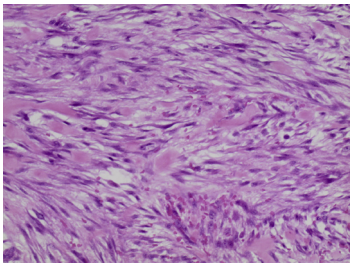

Fig. 5. Haematoxylin and eosin stain at low power demonstrating extravasated erythrocytes intermingled with spindled myofibroblastic/fibroblastic cells.

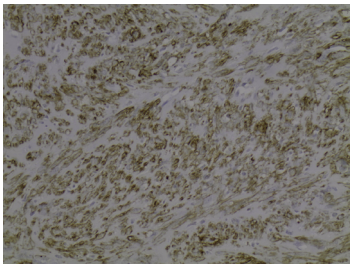

Fig. 6. Immunohistochemistry demonstrating positive staining for smooth muscle actin (α-SMA).

spindled cells for α-SMA, and negative staining for desmin, S100, pancytokeratin AE1/AE3, CD34 and ALK-1 (Fig. 5, Fig. 6). CD34 highlighted blood vessels only.

The patient recovered without any issues at six months, and continues to be monitored for recurrence. She has no functional deficits or signs of recurrence.

3. Discussion

Any rapidly growing facial mass is concerning for neoplasia. The fact that the lesion was well circumscribed and appeared mobile to

adjacent structures suggested a benign neoplasm rather than invasive malignancy. In the area of the left cheek, the potential sources of the lesion could be muscular, neural, connective tissue, or salivary gland (such as that arising from accessory parotid gland).

Nodular fasciitis is a rare and rapidly growing tumour that most commonly affects the upper extremities, especially the volar surface of the elbow [4]. When affecting the head and neck, NF is most commonly oral, but can also affect the face, especially over bony prominences. The true incidence of oral and maxillofacial NF is not known, as it has only been described in a small number of case reports. It may account for between seven and 20 percent of all NF cases, but is usually described in children [1], unlike the case presented here.

When occurring in the head and neck, subcutaneous NF is likely to arise from the superficial layer of the cervical fascia, and presents as a subcutaneous or submucosal well circumscribed lump. Intramuscular and fascial NF may arise from the deep fascial layers, such as in the case presented in this paper.

Nodular fasciitis arises from the fascial layers of muscle, and can extent into the adjacent tissues, or less commonly, into the muscle itself [1]. NF can be categorised into three subtypes; subcutaneous, intramuscular, and fascial. Most cases of NF are superficial, presenting in the subcutaneous tissues. In the rare instance of oral NF, lesions are described as submucosal [1].

3.1. Aetiology and presentation

NF is thought to represent a reactive proliferation of myoblasts or fibroblasts, in reaction to injury [3]. These lesions often may present as rapidly growing well circumscribed masses, or as discrete subcutaneous plaques. They are usually painless [3]. In most cases, lesions present as rapidly growing over months, and may be suspicious for malignancy. In particular, NF lesions can mimic the clinical history and appearance of sarcomas [3]. In most cases, the lesion occurs in an area of chronic irritation, such as over bony prominences.

On computerised tomography (CT) imaging, lesions are enhancing, and have well-defined borders. Intra-muscular lesions may appear less circumscribed, raising concern for malignancy [2]. Magnetic resonance imaging (MRI) of NF reveals hyperintense signal on T2 imaging, and isointensity to muscle on T1. This signal intensity can be non-specific, and NF lesions can look similar to malignant processes such as rhabdomyosarcoma [2].

Due to their aggressive growth pattern, most lesions are treated with surgical excision. Some authors [4] also describe fine needle aspiration (FNA) prior to excision, but this may be inconclusive [2,5,6]. Once excised, NF are unlikely to recur, with recurrence indicative of incomplete excision, or misdiagnosis [1,2,7].

3.2. Histopathology

On histological examination, lesions appear to be un-encapsulated and well circumscribed. The margin between the lesion and surrounding tissue usually demonstrates a pushing front, although direct infiltration has also been described [3]. The lesion is largely cellular, composed of irregular nests and cords of plump spindle shaped fibroblasts and myofibroblasts. These cells demonstrate a high mitotic rate, but exhibit no features of abnormal mitoses. There are no signs of cellular atypia as seen in neoplasia. These cells tend to stain positively for smooth muscle actin (α-SMA), and muscle-specific actin such as HHE-35. Positive staining for actin represents high density of myofibroblasts and presence of muscle fibres. An important characteristic of this lesion is the ground substance, which is typically myxoid and “tissue culture” like. The myxoid degeneration contributes to formation of microcysts. There may be an associated reactive lymphocytic infiltrate, and extravasated erythrocytes. Stains for S-100 and desmin are negative, suggesting that the lesion is not neurogenic (S-100), myogenic (desmin). When compared to sarcoma, NF tend to be smaller on diagnosis (less than 40 mm), sharply demarcated (although infiltrative edges can be seen), myxoid stroma, and scant lymphocytic infiltrate with erythrocytes [3]. Furthermore, sarcomas will exhibit cellular pleomorphism and atypia, which should not be seen in NF.

3.3. Differential diagnosis

Important clinical and histological differential diagnoses of NF include sarcoma [6], fibrosarcoma, fibrous histiocytoma or myofibromatosis [4]. On presentation, NF lesions tend to be smaller than 40 mm, whereas head and neck sarcomas can commonly be more than 50 mm on presentation. Patients may also describe a history of trauma in cases of NF, although this is not always identified. Ultrasound guided fine needle aspiration (FNA) may guide the diagnosis, although in many reported cases, including ours, the FNA result was inconclusive [6]. The issue of misdiagnosis of NF as other malignant pathologies has also been raised by some authors [5] where NF was incorrectly diagnosed as desmoid-type fibromatosis, committing these patients to a more aggressive surgical approach [7].

Initial work up of this lesion suggested schwannoma as a differential diagnosis. Intra-muscular schwannoma has been described as a well-circumscribed mass within the masseter that is non-tender and may feel mobile to the overlying structures. It is well encapsulated with characteristic whirling cells, verocay bodies and Antoni A and B areas. Immunohistochemistry is positive for S-100 [1]. Schwannoma was considered as a probable differential in this case. However, it was important to note that there was no facial nerve dysfunction noted on clinical examination.

Final diagnosis is made on histopathology, specifically with immunohistochemical staining of the excised specimen. For example, leiomyosarcomas stain positive for desmin, whereas NF does not [2]. On confirmation of NF, an excisional biopsy is both diagnostic and therapeutic, with recurrence unlikely. Any reported recurrence should be suspicious for either incomplete excision in the first place, or misdiagnosis of a malignant process [6].

4. Conclusion

Nodular fasciitis is an important differential when assessing rapidly growing solid tumours in the maxillofacial tissues. These lesions can mimic malignancy, and therefore surgical excision is the treatment of choice. Although usually affecting the upper limbs and torso, nodular fasciitis can rarely affect the superficial and deep structures of the oral and maxillofacial region.

The aggressive growth pattern of oral and maxillofacial NF, along with its clinical and histological similarities to sarcoma, make this an important, albeit rare, entity in the head and neck.

Ethical approval

Not applicable

Conflict of interest

None

Acknowledgement

The patient described in this study has given consent for clinical information and photography to be published.

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