

Nodular Fasciitis of the Cheek: A Rare Entity Mimicking Sarcoma

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Keywords

Nodular Fasciitis; Sarcoma; Histology

Introduction

Nodular Fasciitis (NF) is a rare and benign tumor originating from fibroblastic and myofibroblastic tissues. It affects mainly subcutaneous tissue, muscles, and fascia [1].

Although considered traditionally as a reactive lesion rather than a true neoplasm, it is often misdiagnosed as a malignant tumor [2]. We report a case of NF arising in the left cheek mimicking sarcoma.

Clinical Findings

A 30-year-old otherwise healthy man, presented with a 4-month history of an asymptomatic tumor, rapidly growing on the left cheek (Figure 1). There was no history of trauma or systemic complaints. On physical examination, the mass was firm, dome-shaped, painless and adherent to profound structures but not to the overlying skin (Figure 1). Examination was otherwise unremarkable. Magnetic resonance imaging revealed a subcutaneous mass, measuring 20 mm \times 15 mm \times 17 mm arising in the left zygomatic major muscle. The lesion showed hypo-intensity to muscle on T1-weighted images and hyper-intensity on T2-weighted images, as well as contrast enhancement. Based on clinical examination and imaging, we evoked mainly the diagnosis of sarcoma. A skin biopsy was performed. The histopathological study showed a proliferation of spindle cells arranged in a storiform pattern within an alternating myxoid and fibrous stroma (Figure 2). There were 2 mitoses per 10 high power fields (Figure 2). Immunohistochemical staining was diffusely positive for Smooth Muscle Actin (SMA) and negative for CD34 (Figure 2). These findings were compatible with the diagnosis of Nodular Fasciitis (NF). Surgical excision was performed and no relapse was noted on a two year follow up.

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Discussion

Our patient had a challenging clinical presentation of NF misdiagnosed as sarcoma. NF is a rare and benign tumor originating from fibroblastic and myofibroblastic tissues [1]. It affects mainly subcutaneous tissue, muscles and fascia [1]. Although considered traditionally as a reactive lesion rather than a true neoplasm, it is often misdiagnosed as a malignant tumor [2]. In fact, NF is a forgotten entity, which typically presents as a rapidly growing, painless mass rarely exceeding 4 cm



Figure 1: (a) An asymptomatic tumor, rapidly growing on the left cheek; (b) A firm mass, dome-shaped and adherent to profound structures but not to the overlying skin.

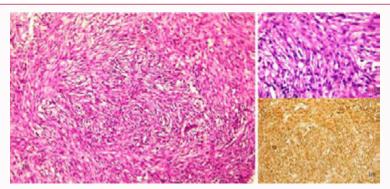


Figure 2: (a) Cellular proliferation showing a storiform pattern (Hematoxylin Eosin x100); (b) Myofibroblastic cells without nuclear atypia or mitosis (Hematoxylin Eosin x400); (c) Tumor cells show diffuse and strong positivity for SMA.

in diameter, with an ovoid shape and well-defined margins. It is most commonly observed on the upper extremities and the trunk, and less frequently on the head and neck as was the case of our patient [2]. The pathogenesis of this benign tumor is poorly understood. Some trigger factors were described in association with NF, mainly trauma and infection [3]. However, NF may be idiopathic, such as this case. NF has also been associated with a somatic MYHP-USP6 gene fusion, suggesting a transient character [1]. Four subtypes of NF are described: Subcutaneous (the most frequent subtype), intramuscular, fascial and unclassified (concerning other locations: Epidermis, knee joint, bladder, prostate, tongue, or parotid gland) [1]. Radiological features can be useful to identify NF. However, it can demonstrate irregular and infiltrative borders, similar to sarcoma, which was the case of our patient. The infiltrative aspect, alongside with rapid growth, cellular and mitotic richness often leads to a misdiagnosis. Histologically, NF can be myxoid, cellular or fibrous. These features can also be seen simultaneously [4]. NF is classically treated with a limited excision, although simple regular observation is possible because spontaneous regression has often been reported. Recurrence is uncommon and no malignant transformation has been reported to date. Awareness of this entity and its benign nature is crucial as misdiagnosis often leads to unnecessary aggressive treatments such as mutilating surgeries [5].

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