



A WORRISOME FAST-GROWING MASS OF THE CHEEK: HOW TO AVOID MISDIAGNOSIS

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Objective: Fibroblastic and myofibroblastic neoplasms of the head and neck encompass a group of rare tumor types with often overlapping clinicopathologic features that range in biologic potential from benign to overtly malignant (1-2). Nodular fasciitis or infiltrative or pseudosarcomatous fasciitis is a benign soft-tissue tumor of fibroblastic/myofibroblastic differentiation. In 7%-20% of the reported cases, the lesion is located at the head-and-neck region (2,3). Intraoral location of the lesion is very unusual and is often confused with sarcomas because of rapid growth, rich cellularity, high mitotic activity and poorly circumscribed nature. The clinical and imaging features of nodular fasciitis are only supportive but not pathognomonic and histopathological examination is mandatory. Histologically, nodular fasciitis is a cellular lesions composed of plump, spindled-to-stellate cells with bland ovoid nuclei arranged in loose fascicles (4,5). More cellular lesions appear vaguely storiform or whorled. The stroma may be predominantly myxoid or collagenous, including areas of keloidal collagen. The cells notably lack nuclear atypia, hyperchromasia, or pleomorphism, but often display a high mitotic rate without atypical forms (4-6). Extravasated erythrocytes and chronic inflammation are common findings. Spindle cells in nodular fasciitis stain positive for vimentin, Bcl-2 and CD34 variably for smooth muscle actin and negative for desmin (1-6). Thus microscopic observation along with ancillary immunohistochemical tests represent the gold standard for avoiding diagnostic pitfalls. We herein describe the clinical and histopathological features of a case of nodular fasciitis of the cheek firstly appeared as a hard swelling of the right masseteric region. Awareness of these entities in the head and neck and knowledge of their clinical behavior can help avoid unnecessarily aggressive treatment for benign tumors such as nodular fasciitis for which spontaneous regression is characteristic.

Methods: We describe the clinico-pathological characteristics of the lesion of a 33 yrs old man who presented with a rapidly growing right cheek intra-masseteric nodular mass of about 2,5 cm which was surgically excised because suspicious for malignancy. We used a combination of immunohistochemical antibodies including S100, Vimentin, BCL2, CD34, smooth muscle actin, desmin, Pan-cytokeratin, MUC-4, MDM-2, β -catenin and Mib-1.

Results: A 33 yrs old man presented to Maxillo-Facial Surgery Department of Azienda Ospedaliera Universitaria delle Marche for a 2 months-lasting hard and painful swelling of the right cheek in the intra-masseteric region. At clinical examination the lesion was slightly nodular and firm without associated lymphadenopathy. Dental assessment and upper aerodigestive tract endoscopy were unremarkable. Initial investigations of an ultrasound scan and fine needle aspiration cytology were inconclusive. The lesion was then excised because suspicious for malignancy. Gross examination showed an almost well-circumscribed multinodular whitish firm tissue of 7,5 cm in diameter. Microscopy revealed discontinuous epithelium overlying variably cellular fibrous stroma with mixed features and diffuse fascicular and storiform arrangement of collagen bundles along with a few areas of infiltrative growth (fig.1-a,b). High power revealed numerous spindle cells displaying occasional mitotic figures, numerous wavy fibers in a mixed and vascularized stroma producing cleft-like spaces (fig. 1-c). Differential diagnoses encompassed spindle cell malignancies such as fibrosarcoma, spindle cell squamous cell carcinoma, leiomyosarcoma and low grade fibromyxoid sarcoma were contemplated and benign spindle cell proliferations such as nodular fasciitis, inflammatory myofibroblastic tumor and solitary fibrous tumor. Immunohistochemical tests revealed immunopositivity for S100, smooth muscle actin (fig.1-d) and vimentin, patchy positivity for BCL2, CD34, while Desmin, pan-cytokeratin, MUC-4, β -catenin and MDM2 were negative. Proliferative index (Mib-1) was approximately of 5%. No dysplasia was observed in the overlying epithelium ruling out squamous cell carcinoma. Overall morphological features along with immunohistochemical results were suggestive of nodular fasciitis.



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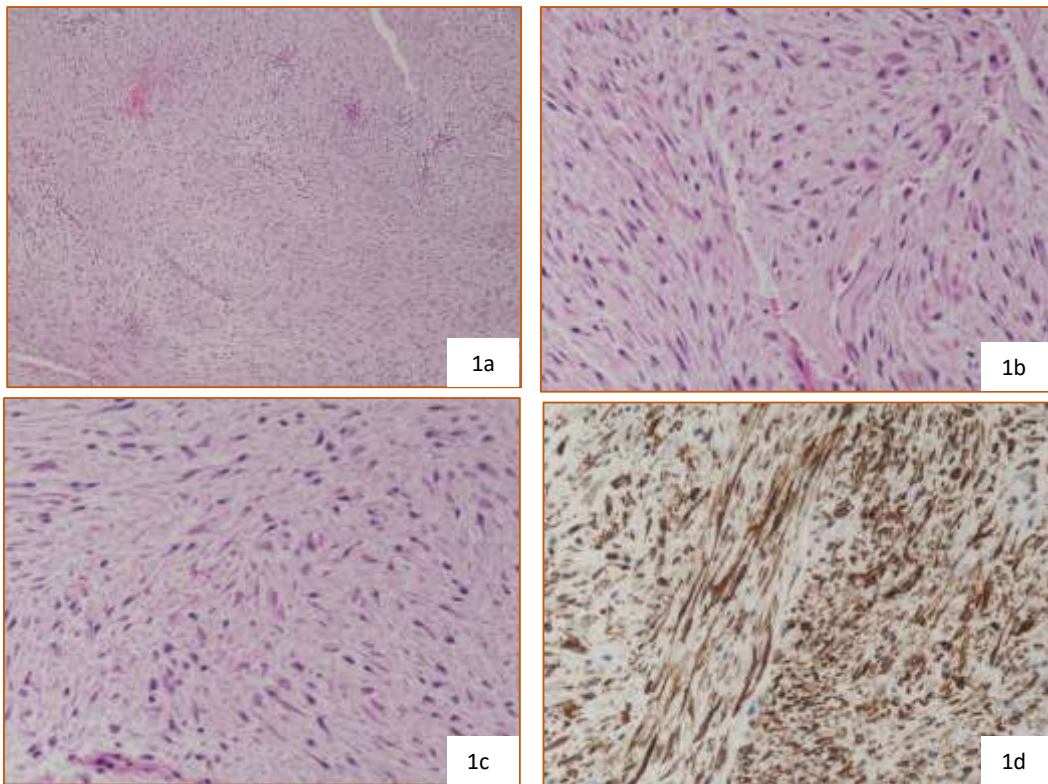


Figure 1: Uniform myofibroblasts are arranged in a storiform pattern, and vascularized stroma producing cleft-like spaces (a,b,c). The cells have a myofibroblastic immunophenotype, with smooth muscle actin positivity (d).

Conclusions: Nodular fasciitis is a benign, rapid proliferation of fibroblasts originating in the deep fascia extending into muscle or subcutaneous tissue. Lesions can occur at any age, but presentation in the third to fifth decade is the most common. Nodular fasciitis in the head and neck, commonly arises on the face or neck, but has also been reported in the oral cavity, orbit, parotid and ear. A small number of cases have also been reported on the masseter such as the present case (7). The aetiology is uncertain, but preceding trauma is reported in 10%-15% of cases.

First described by Konwaller et al., nodular fasciitis is a rare benign self-limiting reactive process of the soft tissues. Although rare, it is the most common soft tissue pseudosarcoma. Its rapid growth, increased cellularity, possibly high mitotic activity and infiltrative borders have resulted in this mesenchymal lesion being misdiagnosed frequently as a sarcoma. The case we presented is strongly interesting not only for the unusual location of the lesion, but mostly because it offers the possibility of stressing the pathological and immunohistochemical features which are mandatory for avoiding a dangerous misdiagnosis of malignancy. The present case offers microscopic and staining details useful for gaining familiarity with the described entity in order to help the pathologists navigate through the treacherous territory of pseudotumorous lesions occurring in the oral and maxillofacial region. Although advanced diagnostic modalities have a considerable role in resolving the interpretative confusion around these lesions, no single investigation is confirmatory. Clinical, radiological, histological correlation and immunohistochemical studies are mandatory for avoiding the diagnostic pitfalls and overtreatment of a patient.

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