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A rare case of nodular fasciitis in cervicofacial region

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OBJECTIVES

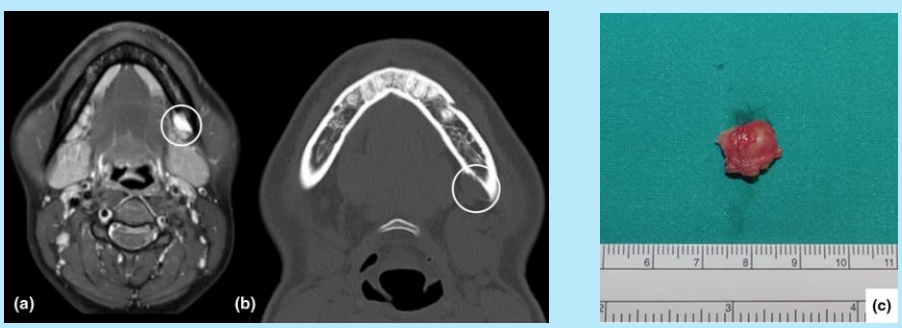
Nodular fasciitis is a reactive process involving a proliferation of fibroblasts. Its pathogenesis is still unknown, although some authors have described trauma as a predisposition to this pathology. A rapid growth, high cellularity and infiltrative borders are the main characteristics of nodular fasciitis.

It is most commonly localized on the upper extremities and trunk, and generally presents between the 2nd and 4th decade of life. the cervicofacial region is the second most frequent location of nodular fasciitis (13–20% of cases, with a greater prevalence in the paediatric population). The mandible is involved in very few cases described in literature. 1,2 The treatment is usually based on surgical excision, and a recurrence of the pathology has been reported in about 2% of cases. In the head and neck region, particularly, a correct balance between surgical safety and invasiveness has to be a priority, assuring at the same time the resolution of the pathology and the best aesthetic and functional results. The aim of our study is to report a rare case of mandibular nodular fasciitis.

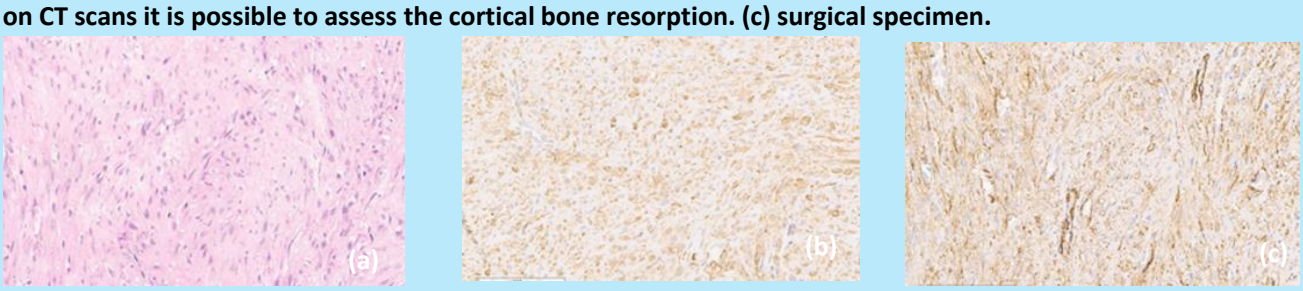
METHODS

The patient, a 16-year-old girl with a round lesion in the submandibular region, was asymptomatic, except for the mandibular swelling. The CT scan identified a round/oval shaped lesion 15×5×7 mm in size, having a solid-nodular aspect (60HU) and involving the bony cortex of the lower medial border of the mandibular body, with cortical resorption. MRI confirmed a 14×7×9 mm lesion, hypointense on T1 images and hyperintense on T2 and STIR with a strong enhancement and lytic alteration of the cortex. FNAC was also performed showing a mesenchymal cell proliferation with no nuclear atypia. Through a 2.5-cm-long incision in the submandibular region, 2 cm from the inferior mandibular edge and assisted by a Hopkins 30° rod lens endoscope, it was possible to access, visualize and perform a complete excision of the lesion, posteriorly to the submandibular gland. Histological examination revealed a nodular, well-circumscribed, mesenchymal cell proliferation, and immunohistochemical staining showed a positivity for smooth-muscle actin and caldesmon and a negativity for S100, CD34, CD68, ALK1 and desmin. These characteristics prompted the diagnosis of a benign myofibroblast neoplastic proliferation, also known as nodular fasciitis.

GRAPHS



Location of the mass in the sub-mylohyoid region (white circle): (a) on MRI scans it appears as a hyperintense mass; (b) on CT scans it is possible to assess the cortical bone resorption. (c) surgical specimen.



Histological specimen: 1) haematoxylin and eosin (x20) showing a cellular neoplasm with a storiform pattern and a variable cellularity. immunohistochemical images show 2)the expression of smooth muscle actin (x20) and 3) caldesmon (x20).

RESULTS AND DISCUSSION

We define nodular fasciitis as a gnoseological identity in its own right, a reactive process involving a proliferation of fibroblasts. Its pathogenesis is still unknown, even though some authors have described trauma as predisposing to this pathology. However, certain recent molecular cytogenetic studies have revealed a recurrent MYH9-USP6 fusion gene, indicating nodular fasciitis as a clonal neoplastic disorder 3. A rapid growth, high cellularity and infiltrative borders are the main characteristics of this condition The typical histological features of nodular fasciitis include haphazardly arranged plump spindle cells within a myxohid stroma. Immunohistochemically, nodular fasciitis is positive for vimentin, smooth muscle actin and muscle-specific actin and it is negative for keratin, CD-34, S100 protein and desmin 4,5. In order to support the diagnosis of nodular fasciitis when accompanied by other tumors with similar histopathological aspects, such as sarcomas and myofibromas, it may be useful to detect the positivity for gene rearrangement of ubiquitin-specific protease 6 (USP-6), present in about 74.4% of nodular fasciitis cases. Our case is in line with the histopathological and immunohistochemical features described in the literature, but presents a rare location.

CONCLUSION

In conclusion, nodular fasciitis is a benign tumour resulting from a reactive process that enhances fibroblast proliferation. The head and neck region is involved in about 25% of cases with a slight male prevalence (M:F = 1.5:1). Nodular fasciitis can be diagnosed with CT scans and histological examinations, but the detection of a translocation of USP-6 can be helpful to distinguish nodular fasciitis from malignant neoplasms which share an overlapping histology.

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