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A rare case report of a typical variant ossifying fibromyxoid tumor (OFMT), located in the retroauricular perimastoid region

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ABSTRACT

INTRODUCTION: Ossifying fibromixoid tumor of soft parts (OFMT) is a rare soft tissue and bone tumor. In its classic form, is considered benign, nevertheless aggressive clinical behaviour tumors with a different cytoarchitectural features of a malignant variant, have been described.The classification contains "typical", "atypical" and "malignant" variants.
METHODS: A CT ear scan without contrast enhancement was carried out (October 2015), with coronal, sagittal and 3D reconstructions. It was decided the removal of the neoformation with a simple dissection.
RESULTS: In this report, we present a case of a typical variant OFMT localized in the retroauricular perimastoid region, with mastoid bone cortex not involved.
CONCLUSIONS: Considering the extreme rarity of this "enigmatic" tumor which displays an uncertain line of differentiation, renders the differential diagnosis a true challenge.

1. Introduction

Ossifying fibromyxoid tumor of soft parts (OFMT) is a rare soft tissue and bone tumor, described for the first time in 1989 by Enzinger et al. [1], which prevalently affects the subcutaneus tissue of extremities and trunk, although other body sites have been reported, such as the mediastinum [2] and neck [3]. It has an uncertain histopathogenesis and is characterized by a peripheral bone shell embedded in a fibromyxoid matrix, which tends to have a rough, small round cell proliferation with a very low mitotic activity. In its classic form, it is considered benign, nevertheless aggressive clinical behaviour tumors with a different cytoarchitectural features of a malignant variant, have been described[4–6]. Folpe and Weiss proposed a classification with "typical", "atypical" and "malignant" variants [4] and affects males to females at a ratio of approximately 1.5:1, with a median age of 50 years [7]. In this report, we present a case of a typical variant OFMT, localized in the retroauricular perimastoid region, in line with the SCARE criteria [8].

2. Case report and results

A 31 years old patient presented in the ENT department, on November 2015, reporting a slowly growing mass behind his right ear. The physical examination revealed a solid mass, painless to palpation, absence of ulcers or any other skin lesions, with the dimensions of approximately 2 cm, in the right perimastoid region. Otoscopy was normal, also audiometry did not revealed any abnormalities, such as conductive hearing loss. The medical history hasn't revealed any other co-morbidities and/or cronic diseases. A CT ear scan without contrast enhancement was carried out (October 2015), with coronal, sagittal and 3D reconstructions (Fig. 1). The exam revealed a right perimastoid mass formation ($2.2 \times 1.2 \times 2$ cm) with an oval morphology, extended right to the back of the external ear canal, well cleaved and separated from the mastoid bone cortex, characterized by the predominant calcific component. The mastoid cells had a normal pneumatization. There were no obvious abnormalities of the middle and internal ear structures bilaterally. It was decided the removal of the neoformation with a simple dissection. A written informed consent for the use of anonymous data, eventually for a future study, with the informed consent for the biopsy/operation, were obtained from the patient. During the operation it was confirmed that the mastoid bone cortex wasn't involved.

On histological examination, in all the preparations prepared with hematoxylin-eosin staining, the sections are represented by a

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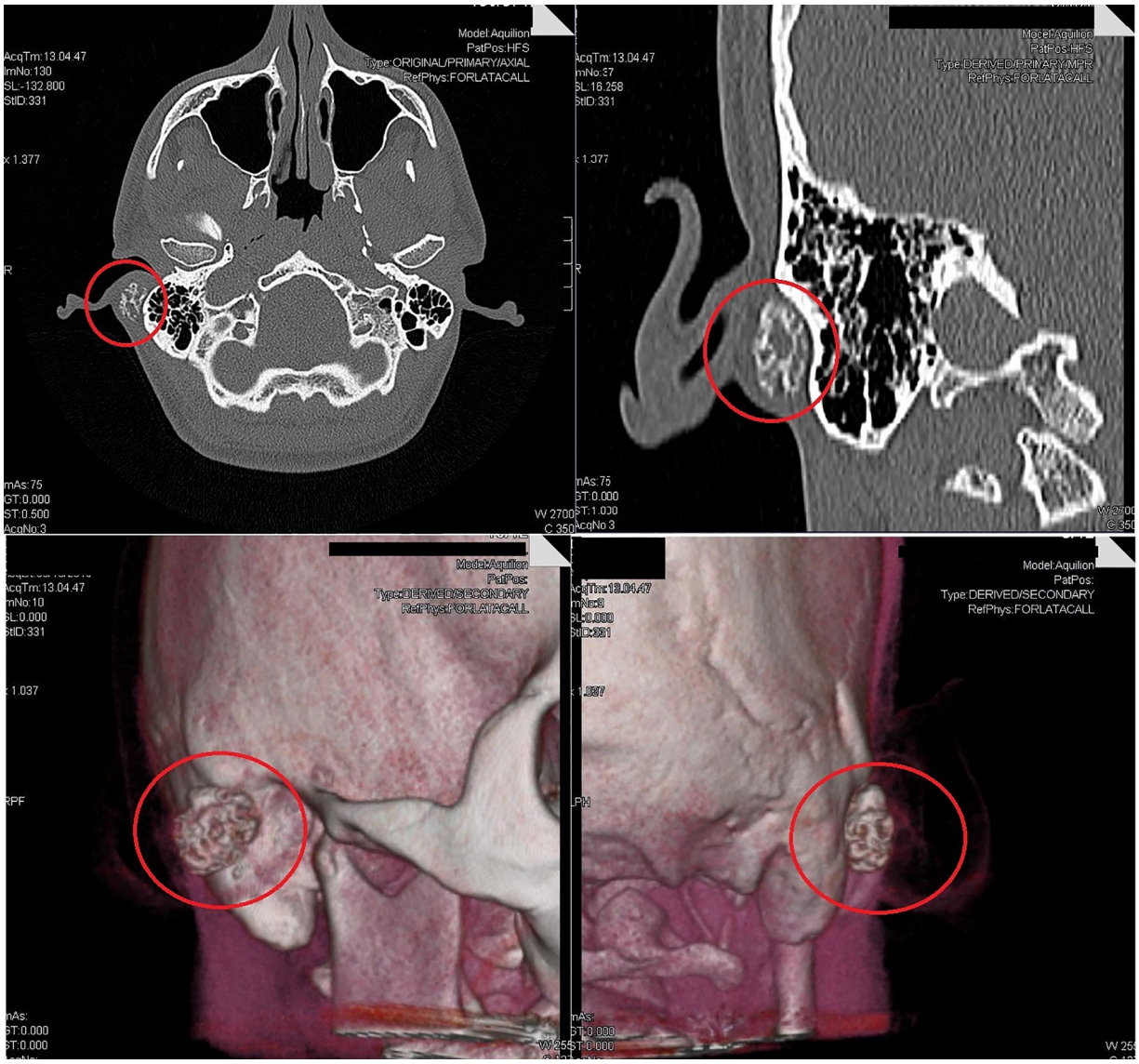

Fig. 1. CT ear scan with coronal, sagittal and 3D reconstructions, with red circles pointing the location and the extension of the neoformation.

nodular lesion composed by a proliferation of rounded or fusiform cellular elements with leptochromatic nucleus. The cytoplasm is described as a poorly eosinophilic, arranged to form solid nests and cords immersed in abundant collagenous and/or myxoid stroma, interposed in a mature irregularly shaped trabecular bone, mainly located on the periphery of the lesion. Also spots of calcification and some small dilated vessels were present. There were absence of mitosis and areas of necrosis(Fig. 2). The above cellular elements were positive with immunohistochemical staining for S100 protein and vimentin and negative with immunohistochemical staining for Pan-CK, CK7, CK20 and EMA. The proliferation index assessed with Ki-67, is approximately is 1%. Two years after the diagnosis and the removal of the tumor there hasn't been any relapse or metastasis, in accordance with the prognosis of a typical variant OFMT. The patient is in good condition, without any symptoms or pathological conditions related to the OFMT.

3. Discussion

OFMT is a soft tissue tumor of uncertain histogenesis, which occurs more often in adults, with a higher incidence in males than females. Differential diagnosis includes myositis ossificans, ossifying hematoma, tumoral calcinosis, extraskeletal chondroma, low-grade fibromyxoid sarcoma, synovial sarcoma and extraskeletal or parosteal osteosarcoma [9].

Typically, OFMT presents as a slow-growing benign mass, nevertheless Graham et al. [6] confirmed the existence of malignant OFMT using immunohistochemistry and gene expression profiling, suggesting also that this tumor has a scrambled phenotype. Miettinen M et al. [7] evaluating malignancy parameters found that an increased mitotic activity, is a risk factor of local recurrences, whilst tumor size and necrosis are not significant risk factors. Folpe AL, Weiss SW [4] demonstrated that hypercellularity, nuclear atypia and high mitotic index activity (>2/50 HPFs), represent risk factors for aggressive behaviour with local recurrences and metastases, characteristics of the malignant variant, when low nuclear grade, low cellularity and a mitotic rate of <2/50 HPF,are the features of a typical variant. Cases that deviate from typical OFMT but do not meet malignant criteria can be designated as atypical OFMT. In immunohistochemistry the positivity expression level of S-100 protein tends to decrease as the tumor grade increases: 88% for typical cases, 75% for atypical cases, and 42% for malignant cases [6].

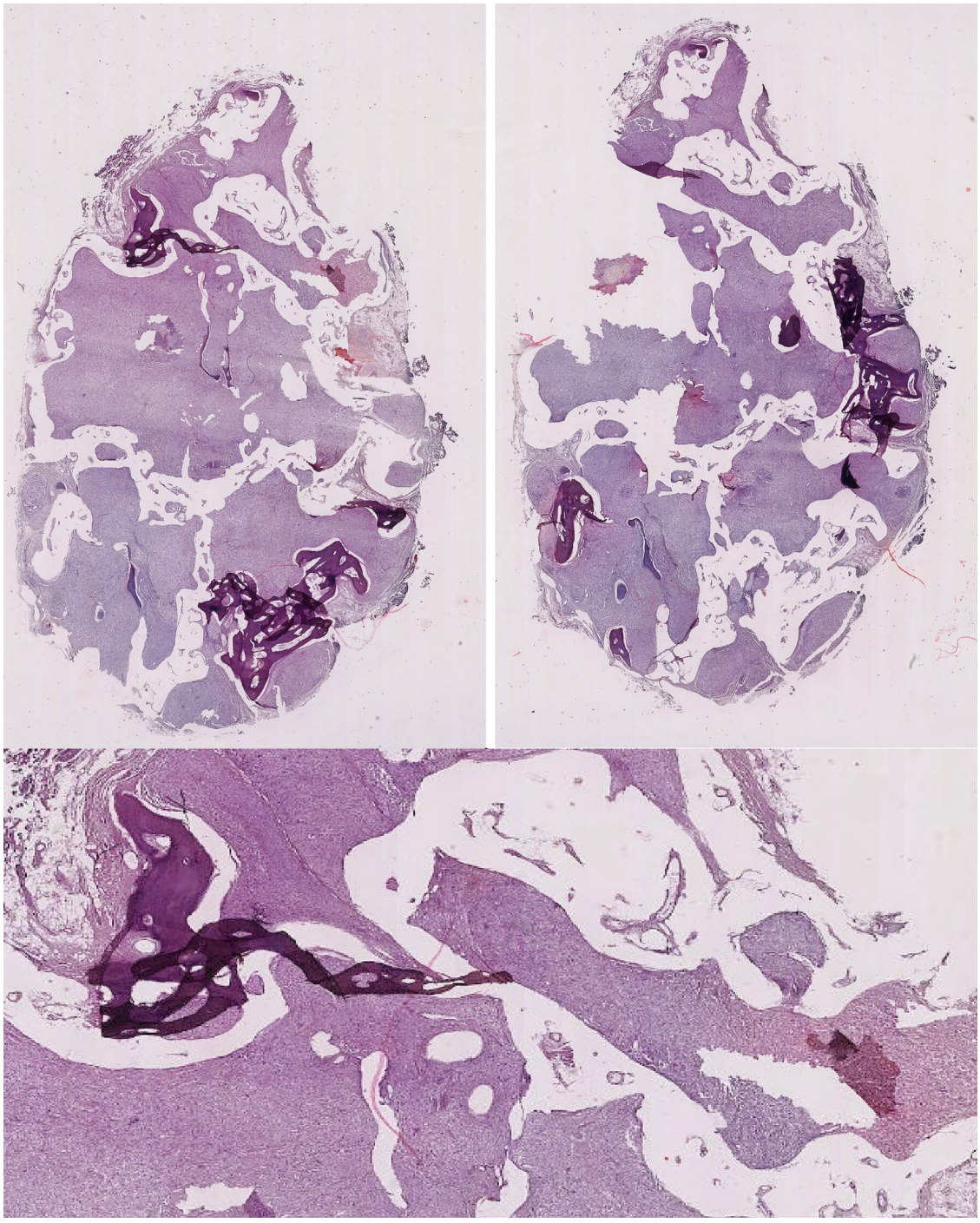

Fig. 2. Histologic preparation: Fusiform cellular elements, eosinophilic cytoplasm. Note the mature irregularly shaped trabecular bone, mainly on the periphery of the lesion.

Involvement of the head and neck area is infrequent (10% to 15%) and intraoral presentation is very rare [10,11]. Nevertheless, tumor locations such as the oral cavity, mandibular gingival, tongue, etmoid sinus and other sites of the head and neck region have been described in the literature [11].

4. Conclusion

In conclusion, we present a case of a typical variant OFMT that occurred in an extremely unusual site, in the retroauricular perimastoid region. Considering that it is extremely rare, and the possibility of local metastasis especially for the malignant type, we believe that more studies are required for this "enigmatic" tumor which displays an uncertain line of differentiation, rendering the differential diagnosis a true challenge.

Conflict of interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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Ethical approval

In Italy Ethics Committee stands for Institutional Review Board. This should be applied indeed for patients, but for studies such as clinical trials, invasive and/or experimental therapies. Since our paper is a case report there wasn't any Ethics Committee approval. Instead there was a written and signed consent from the patient for the operation and for the use of anonymous data eventually for research.

Consent

There was a written and signed consent from the patient for the operation and for the use of anonymous data eventually for research.

Author contribution

Dr. Theodoros Varakliotis: Design, acquisition and analysis of data, drafting and revision of the manuscript,
Prof. Gianluca Bellocchi: Supervision, revision of the manuscript
Prof. Alberto Eibenstein: Supervision, revision of the manuscript
Dr. Gilberto Acquaviva: technical support
Dr. Francesco Casorati: technical support, collection of the data

Guarantor

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