Case report

Desmoplastic fibroma of the mandible

Raffaele Averna[1], Massimo De Filippo[1], Silvano Ferrari[2], Ermanno Bacchini[3], Cristina Rossi[1]

[1] Department of Clinical Sciences, Unit of Radiological Sciences, University of Parma, Parma Hospital, Parma, Italy; [2] Department of Maxillo-Facial Surgery, University of Parma, Parma Hospital, Parma, Italy; [3] Department of Clinical Sciences, Section of Radiological Sciences, Pediatric Division, University of Parma, Parma Hospital, Parma, Italy

Abstract. We report the imaging findings of a desmoplastic fibroma (DF) of the mandible in a 3 years old girl. DF of bone is a rare, no-metastasizing but locally aggressive tumor. Hypercellularity, nuclear pleomorphism, mitotic activity, and traces of odontogenic epithelium and bony tissue are absent. US exam showed a highly vascular and well delimited mass, with no necrotic/hemorrhagic areas. It appeared as a well-defined osteolytic region in RX and a multiloculated, hypodense mass, with no periosteal reaction signs, in CT scans. MRI showed hypointensity in T1w TSE sequence and hyperintensity both in T1w TSE SPIR and T2w ones with no restriction of the "apparent diffusion coefficient" (ADC). In conclusion, remaining histology the gold standard for the DF diagnosis, imaging features may strongly suggest it. (www.actabiomedica.it)

Key words: desmoplastic fibroma, bone mass, pediatric benign neoplasm

Abbreviations

DF:	desmoplastic fibroma
US:	ultrasound or ultrasonography
CT:	computed tomography
MR:	magnetic resonance
LGCSO:	low grade central osteosarcoma
TSE:	turbo spin echo
SPIR:	spectral presaturation by inversion recovery
ADC:	apparent diffusion coefficient
MPR:	multiplanar reconstructions
FNAC:	fine-needle aspiration cytology
ABC:	aneurysmal bone cyst
OPT:	orthopantomography

Introduction

Desmoplastic fibroma (DF) of bone was firstly described by Jaffe HL in 1958 (1). DF is a rare tumor with a no-metastasizing but locally aggressive behavior and a reported incidence of 0.1% of all bone tumors and of 0.3% of all benign bone tumors (2).

The mandible is the most involved bony segment of the body (40%), followed by long bones and pelvis (3); in a review by Ikeshima A. et al., upon 69 cases of DF of the mandible, 57 occurred in the jawbone and 11 in the maxilla, showing that the jawbone is the structure where this neoplasm more frequently grows (4). The ramus, angle and molar area are the most affected sites; about long bones, the metadiaphysis are the most common interested ones (3).

Symptoms include: local painful swelling, chewing/swallowing difficulty and, rarely, loss of teeth. Signs can be represented by facial asymmetry and abnormal teeth position.

In literature we found case reports with patients of any age, since 6 months until 70 years old and over, but the first, the second and the third decades of life are the most represented (3).

There is not an unanimous agreement in considering sex as a risk factor. Most of the authors affirm that there is no difference between male and female,

while other ones indicate a slight predominance of men: in 2005, Frick M.A. et al. reported a male-female ratio of 1.5:1, in a series of 93 patients (2).

DF is considered the intraosseous counterpart of the soft-tissue desmoid tumor (aggressive fibromatosis of soft tissue).

Histology represents the gold standard for its diagnosis because DF hasn't got any pathognomonic imaging features.

Hypercellularity, nuclear pleomorphism, mitotic activity, and any traces of odontogenic epithelium and bony tissue are absent, differentiating this lesion specially from ameloblastoma, odontogenic keratocyst, solid variant of aneurysmal bone cyst, fibrosarcoma and low grade central osteosarcoma (LGCOS).

Cause of its high recurrence rate, a radical excision surgery should be preferred in the treatment. Curretage is in fact accompanied by a 72% recurrence rate, versus the 17% of surgical resection (5).

Case report

A 3 years old girl suffering of a swelling on the right side of the mandible, appeared some months earlier, was referred to our pediatric radiology department because of its size improvement. In a different institute she received the diagnosis of laterocervical lymphadenomegaly but, after two months of ineffective antibiotic therapy, her parents decided to bring the child to our attention.

She showed a clear face asymmetry with a quite big swelling on the right side of the mandible. The mother reported that the child had no relevant symptoms until few days earlier when an ingravescent difficulty in swallowing occurred. Parents also referred that some weeks before the child fell and hurts itself just in the right jowl, but any relationship between a trauma and the DF development is still completely unproven.

On tapping examination the swelling was firm and appeared to be fixed to the mandible. There were no alterations of the skin, but dental examination showed the lateral dislocation of the second premolar tooth.

US exam, also with color Doppler technique, showed a solid, highly vascular and well delimited mass, with a dishomogeneous echogenicity structure but no necrotic or hemorrhagic areas. We realized that it could not be expression of laterocervical lymphadenomegaly cause of the lack of any recognizable lymphnodal echographic features.

To check the bone conditions, we performed a RX examination with three different views (anteroposterior, latero-lateral, oblique) which evidenced a large and well-defined osteolytic region in the context of the mandible structure, with the postero-lateral dislocation of the first included molar teeth. The lesion showed a multiloculated architecture with some bony trabeculas going through it. There were no periosteal reaction signs (fig. 1a,b,c).

In order to better evaluate its nature, we decided to subject the child to MR examination, with and without contrast medium, completing it with MR diffusion sequences. It showed a 56 x 42 x 49 mm solid mass arising from the medial side of the right hemimandible, causing the remodeling of the bone. There were some hypertrophic vascular structures and several enlarged reactive laterocervical lymphnodes. The lesion exerted a mass effect to the next anatomical structures causing the left dislocation of the tongue with consequent difficulty in chewing and swallowing. It appeared to be hypointense in T1w TSE sequence and hyperintense both in contrast-enhanced T1w TSE SPIR and T2w ones (fig. 2a,b,c). We didn't find any "apparent diffusion coefficient" (ADC) restriction in the MR diffusion sequences (fig. 3).

On the same day, the child underwent to CT examination to evaluate the bone involvement. After multiplanar reconstructions (MPR) we obtained images in the three orthogonal planes showing a well-defined, multiloculated and hypodense mass, subverting the structure of the ramus of the jaw, growing and pushing from inside to outside. It was possible to see the same included first molar teeth and trabeculas already described in RX exam. There were some bony erosion signs but no periosteal reaction, mineralized matrix or cortical infiltration ones (fig. 4).

Being no signs of bone or soft tissues infiltration and no restriction of the ADC, we concluded that the lesion was a benign but locally aggressive neoplasm without inclination to malignity.

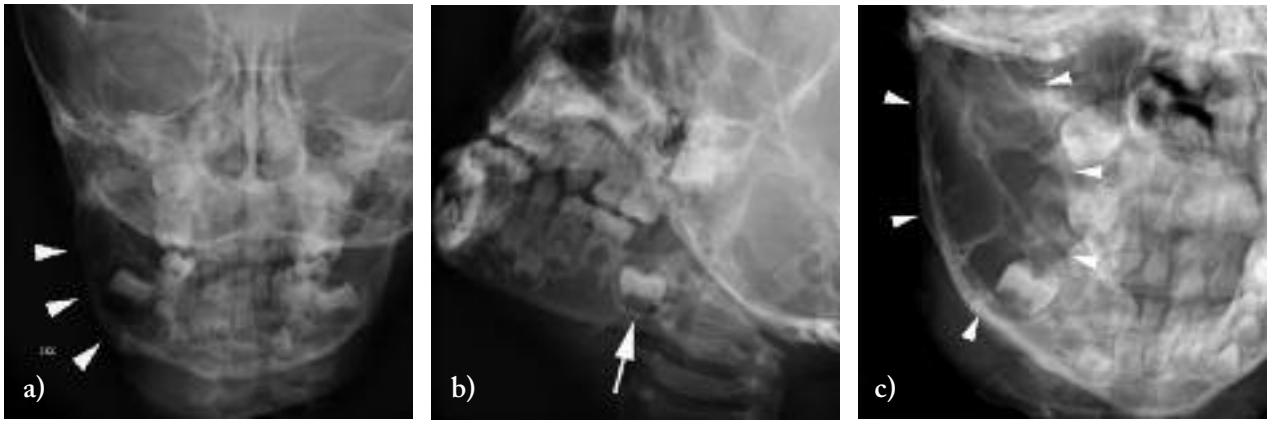

Figure 1. RX images in antero-posterior (a), latero-lateral (b) and oblique (c) view with a large, multiloculated and well-defined osteolytic region in the context of the mandible structure (arrowheads), with the postero-lateral dislocation of the first included molar teeth (arrow). There are no periosteal reaction signs.

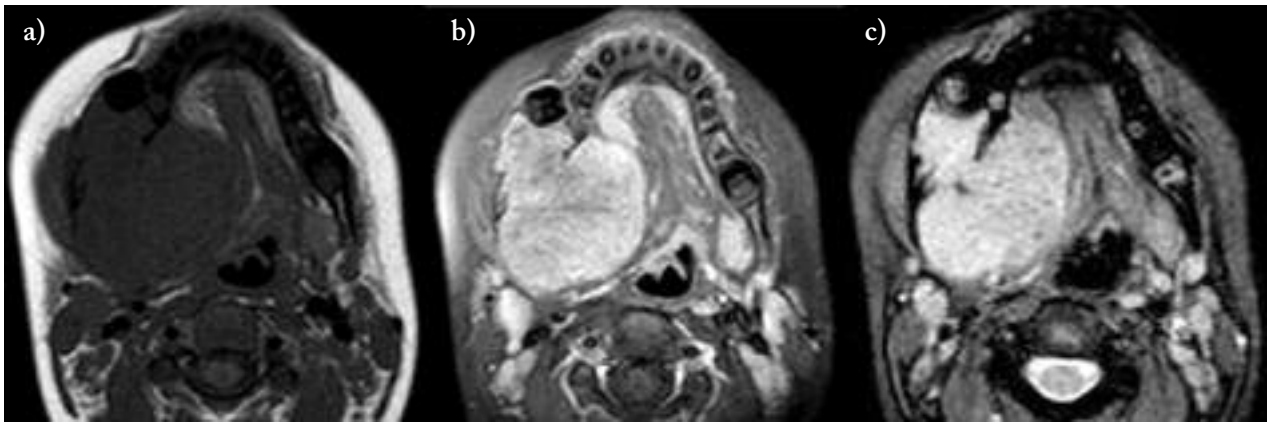

Figure 2. Axial T1w TSE sequence (a) showing the hypointensity of the mass. Axial T1w TSE SPIR sequence (b) and axial T2w one (c) with the high intensity signal of the lesion.

A FNAB was suggestive of DF. The child underwent to maxillo-facial surgery. Surgeons performed, in the same session, a right hemimandibulectomy and the reconstruction of the sick jawbone. They got it removing a rib from the child's chest and inserting it in place of the missing hemimandible.

The diagnosis was confirmed by the following histological examination.

In the differential diagnosis, DF must be consider one of the main suspected neoplasms when there is an occurrence of any lytic, cystic and locally aggressive intraosseous lesion in childhood, especially when the mandible is the involved bony segment.

Discussion

Up to the present, less than 200 of DF cases have been reported in literature, and fewer than 100 regarding the mandible. Moreover, a very little part of them describes its US and MR features.

DF is hystologically formed by interlaced bundles and whorled aggregates of densely fibroblastic collagenous matrix, containing uniform spindle and elongated fibroblasts. It's characterized by low cellularity with some possible foci of hypercellularity. No cytologic atypia or mitotic figures neither osseous matrix are observable.

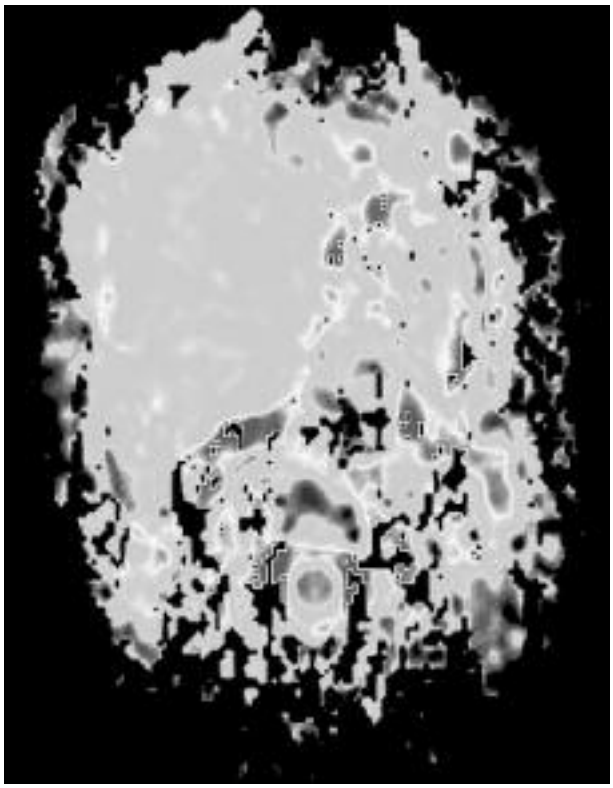

Figure 3. MR diffusion sequence with no restriction of the ADC, confirming the low cellularity of the DF.

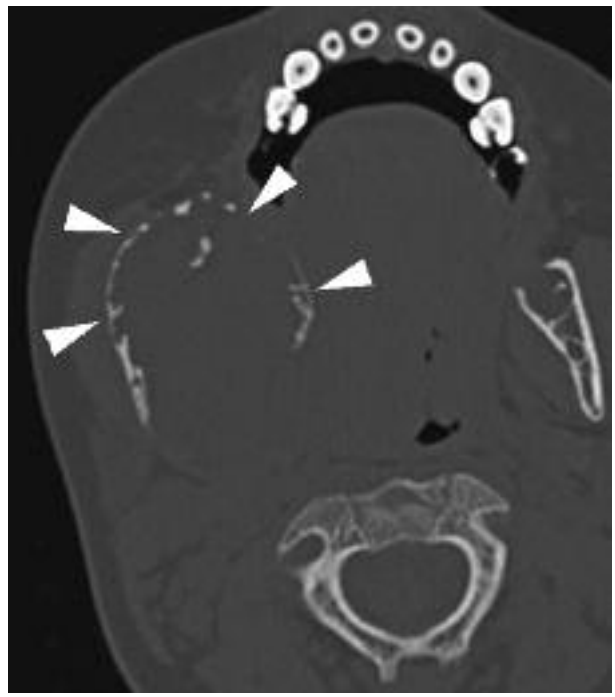

Figure 4. CT scans in axial view. A multiloculated and hypodense mass subverts the structure of the mandible ramus (arrowheads). There are some bony erosion signs but no periosteal reaction, mineralized matrix or cortical infiltration ones.

The DF echographically appears as a solid and highly vascular mass with a dishomogeneous echoic structure. Differential diagnosis must includes: rhabdomyosarcoma, intraosseous hemangioma (cavernous type) and solid variant of aneurysmal bone cyst.

Radiographically we can observe a radiolucent, osteolytic and multiloculated lesion more often than uniloculated one, without any periosteal reaction signs (fig.1 a,b,c).

Differential diagnosis includes osteomyelitis, ameloblastoma, non ossifying fibroma, giant cell reparative granuloma of bone, intraosseous hemangioma, histiocytosis X, odontogenic keratocyst and, more rarely, primary malignant lesions such as osteoclastoma, low grade fibrosarcoma and low grade central osteosarcoma (LGCOS). The latter in particular is often indistinguishable from DF having a low vocation to the cortex and soft tissues involvement that tends to mask its aggressive character. LGCOS represents the 1-2% of all osteosarcomas and concerns the long bones in the 80% of cases. Its prognosis is generally good, including 5- and 10-year survivals of 90% and 85% respectively.

However, recurrence is not an uncommon event, particularly in patients treated with curettage or local excision.

MR findings of DF are not often described in literature. The greater part of the authors, i.e. Frick MA et al. and Kendi TK et al., report a substantial hypoisodensity both in T1w and T2w sequences (2, 6). Our case instead presents hypointensity in T1w TSE and hyperintensity in contrast-enhanced T1w TSE SPIR sequences and in T2w ones, resembling the findings reported in the papers by Kong KY et al. (2000) and by Urresola A. et al. (2007) (fig. 2a,b,c) (7, 8). Everyone instead agrees about the intense contrast enhancement of this neoplasm. In no report we found any reference to the MR diffusion sequences features.

Diffusion is a physical process based on the random movement of water molecules, known as Brown-

ian movement. Diffusion-weighted MR imaging is a relatively new sequence that provides information on biophysical properties of tissues as density, cell organisation and microstructure, which influence the diffusion of water molecules. In biological tissues, diffusion is not truly random because tissue has structure, therefore water diffusion is referred to as "apparent diffusion". A low ADC corresponds to high signal intensity (restricted diffusion), and a high ADC to low signal intensity on diffusion-weighted images. Our findings about this parameter (ADC mean value = $1.42\pm0.37 \times 10^{-3}$ mm^2/s) indicate no restriction of the "apparent diffusion coefficient" (ADC), confirming the substantial low cellularity of this kind of tumor (fig. 3).

CT examination allows to verify the bony involvement entity. DF appears as a hypodense, well-delimited mass and it's not unusual to find teeth jailed inside it. Like we find in RX images, also in CT scans we can mainly see a multiloculated lesion growing inside the bone structure and giving it a sort of "internal explosion" aspect (fig. 4). No malignancy signs, such as periosteal reaction, osteolisys or soft tissue infiltration, are present.

Even if endowed with high accuracy, we must say that these signs haven't got absolute specificity and sensitivity. They can be absent in central malignant bone neoplasms, if the bone cortex hasn't been still involved, or in case of low malignant potential tumor, like the LGCOS is. We can instead find those signs also in other pathologic entities, such as osteomyelitis and solid variant of aneurysmal bone cyst (ABC).

Anyhow, the presence of periosteal reaction, osteolisys or soft tissue infiltration, in the greater part of cases demonstrates the malignant nature of the bony lesion and CT is the most accurate imaging diagnostic procedure to define it.

In conclusion, remaining histology the gold standard for the diagnosis of desmoplastic fibroma, imaging findings and MRI and CT ones in particular, can at any rate strongly address to its identification.

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Correspondence: Massimo De Filippo, MD
Department of Clinical Sciences, Unit of Radiological Sciences
University of Parma, Parma Hospital
Via Gramsci, 14
43100 Parma, Italy
Tel. 0039-521-703660
Fax 0039-521-703491
E-mail: massimo.defilippo@unipr.it