Case report

Juvenile trabecular ossifying fibroma in an 12 year old patient, a case report

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Keywords

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Abstract

Juvenile ossifying fibroma (JOF) is one of the rarest entities within the very heterogeneous and large group of benign fibro-osseous tumors. It is often diagnosed in the first two decades of life, is usually asymptomatic, and affects males and females equally. JOF commonly arises in the mandibular or maxillary region and has a particularly aggressive behavior, characterized by rapid growth and a high risk of recurrence (estimated at 20-50%). Surgical enucleation, followed by curettage and/or osteotomy remains the mainstay of treatment.

Case Report

A 12-year-old boy presented for investigation of a painful mandibular mass noted after minor trauma to the mandible. The patient was apyretic and there was no evidence of infection. His past medical history was uneventful. Physical examination confirmed an inferior-lateral right mandibular mass that was firm and nonmobile, without skin lesions. Full mouth opening was prevented by the lesion, thus solid food intake was compromised.

Blood tests showed no evidence of inflammation. Serological screening was negative. A CT scan of the mandible showed an encapsulated formation of approximately 5 cm in width, located within the right ascending branch of the lower mandible, facing the dental root 48. MRI showed no evidence of bone destruction. An FDG PET-CT showed hyperavidity of the lesion with a suspicious adenopathy in level IIa (upper jugular nodes), homolaterally (Figure 1 and 2).

A biopsy was performed and revealed a tumoral structure with osteogenic differentiation and lamellar osteoid matrix, amidst a disrupted osteoblastic and connective structure. Histology was consistent with a juvenile trabecular ossifying fibroma.

Conservative surgical endobuccal resection was performed, followed by 6 weeks of maxillomandibular block to optimize bone consolidation (Figure 3).

Full mandibular mobility and masticatory forces were regained after removal of the consolidation block.

However, the 12 month follow-up MRI showed clear signs of relapse in the lower right mandible. The CT scan confirmed the presence of multiple tumoral lesions located within the right posterior mandible and possibly the coronoid process.

Considering the extent of the lesion and the short remission period, curettage was not considered an appropriate option. A radical resection of the posterior mandible, including the condylar process, is planned. Reconstruction will be performed with a fibular graft.

Discussion

Juvenile ossifying fibroma (JOF) is a rare benign entity belonging to the group of fibro-osseous tumors. JOF has a clinically more aggressive nature and a high recurrence rate compared to a conventional ossifying fibroma generally seen in adults (1,2). The underlying molecular mechanisms remain largely unknown (1,3).

There are two histological subtypes of JOF: the trabecular and the psammomatoid JOF (1-4).

The trabecular subtype is characterized by a cellular fibrous stroma composed of spindled fibroblast cells with a lamellar osteoid matrix without osteoblastic rimming and immature bony trabeculae surrounded by plump osteoblasts (1,4,5). Trabecular JOF affects the craniofacial bones, predominantly the jaw (1,5).

In contrast, the psammomatoid subtype contains cellular fibroblastic stroma with curved and spherical ossicles (1,2,4,5). Psammomatoid JOF more commonly involves the paranasal sinuses and the periorbital region (2,5).

Figure 1: Head and neck MRI T1 with fat suppression and contrast (Gadolinium).

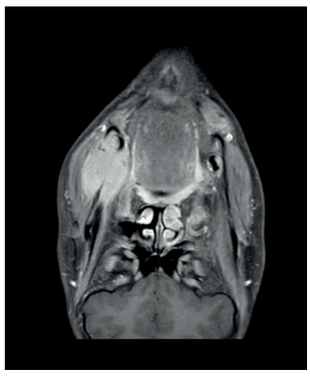
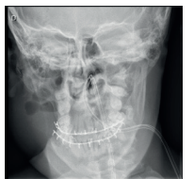


Figure 2: FDG PET-CT.

Figure 3: Post Surgery X Ray.



Trabecular JOF is generally found in younger patients (< 15 years) compared to psammomatoid JOF (mean age 16-33 years) (2,3,5,6). There doesn't seem to be a gender predilection, although some studies describe a male:female ratio of 3:2 (1-4).

Clinical presentation is silent in most cases. Dental displacement and indolent swelling are often described (1,2,3,5). Nasal obstruction, ptosis or exophthalmos may be found in paranasal or periorbital locations.

In some cases, previous trauma is associated with JOF (1).

Although JOF is often locally aggressive and prone to recurrence (estimated 20-50% recurrence rate), malignant transformation to sarcoma has not been reported so far (2,3). Even though the lesions are usually well demarcated, their rapid growth and its osteolytic nature with cortical thinning may mimic malignant entities such as sarcoma, hematopoietic neoplasms, secondary metastasis or odontogenic tumors (3,6,7).

The lesions often present as unilocular mixed lesions, with radiolucent and radiopaque entities (1,2). The cortical outline remains intact, although cortical thinning may be seen (3,5).

Clinical, histological and radiological features assist the clinician in differentiating a malignant lesion from a benign lesion. An overview of the differential diagnosis can be found in the Appendix.

There is no consensus regarding optimal treatment. Enucleation and curettage with peripheral ostectomy remains the main first-line treatment strategy (1,6). Second line treatment consists of hemimaxillectomy and peripheral resection (8). In some cases bone grafting is required for reconstruction. This procedure is particularly challenging in the pediatric population, as bone growth is still ongoing.

In most studies, there doesn't seem to be a difference in outcome between limited or extensive surgery (1). However, some studies have reported lower recurrence rates after complete resection (5,8). Because osteotomy has a much higher morbidity rate, it remains the second treatment choice (5,8).

Patients often face a long rehabilitation process with prolonged maxillomandibular block, esthetic challenges and articular mandibular dysfunction leading to feeding difficulties.

Conclusion

JOF is a benign fibro-osseous tumor without potential of malignant transformation. Still, its rarity and its silent presentation can make diagnosis challenging. Differential diagnosis with odontogenic tumors, sarcoma, hematopoietic neoplasms, Langerhans cell histiocytosis or secondary metastasis is important. Surgical resection is the mainstay of treatment, although harboring its own challenges, such as aggressive surgery, sometimes requiring bone grafting, an high local recurrence rates. These features contribute to increase JOF's potential morbidity. Long-term follow-up is strongly recommended (1).

Conflict of interest

The authors have no conflict of interest to declare with regard to the subject discussed in this manuscript.

APPENDIX: Recapitulative chart of the differential diagnosis.

	Sex Ratio	Age	Radiological features	Histological features	Treatment	Prognosis
Juvenile ossifying fibroma	No gender predilection	< 15 years	Osteolytic nature with cortical thinning	Trabecular or psammomatoïd type	Enucleation/curettage or peripheral ostectomy	High recurrence rate: 20-50%
Ossifying Fibroma	9:5 female:male ratio	Mostly adults, 20-40 years	Mixed radiolucent / radiopaque lesion	Trabeculae of woven bone lined by active osteoblasts	Enucleation/curettage or peripheral ostectomy	Very rare recurrence
Langerhans cell histiocytosis	2:3 male:female ratio	Children < 15 years, often 1-4 years	Osteolytic lesion with irregular margin	Clonal proliferation of Langerhans Cells	Surgical curettage / systemic chemotherapy / corticosteroids	- Depending on initial site - Unisystemic or multisystemic lesion
Osteoblastoma	Male predilection	Adults, often 20-30 years	Mixed radiopaque and radiolucent lesion	Bony trabeculae lined by osteoblasts	Surgical Excision (+ Radiotherapy)	Recurrence rate: up to 20%
Osteosarcoma	Male predilection	Adults, often 30-40 years	Osteolytic and osteogenic nature witch cortical destruction	Immature osteoid formation: proliferation of neoplastic mesenchymal cells with spindle-shaped, epithelioid, or plasmacytoid appearance	- Ablative surgery - Adjuvant chemotherapy - Adjuvant radiotherapy	5 year survival rate of 65%
Odontogenic tumors	9:7 male:female ratio	Mean age 11.6 years	Radiolucent lesion	Fibromyxoid tissue with variable cellularity surrounded by a cuboidal to columnar odontogenic epithelium	Excision / enucleation	No reports of recurrence
Hodgkin Lymphoma	Male preponderance 3:1	Peak in 4th decade, but affects also children	FDG/PET-CT MRI with increased T2 signal	Presence of Hodgkin/ Reed Sternberg cells	Radiotherapy/chemotherapy	Ten year survival 80-99%
Non Hodgkin Lymphoma	Male predilection	Average age of 65 years (Several types mostly in children like Burkitt's Lymphoma)	FDG/PET-CT MRI with increased T2 signal	Absence of Hodgkin/ Reed Sternberg cells	Radiotherapy/ chemoimmunotherapy	5 year survival rate of 72,7%
Fibrous Dysplasia	Slight female predilection	Mean age 20 years	CT Scan: - Radiodense lesion with ground glass appearance - Radiolucent/cystic lesion	Cellular fibrous connective tissue with irregular bony trabeculae	- Surgical resection - Bisphosphonates - « Wait and see »	Recurrence rate: up to 84%

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