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Desmoplastic fibroma of the mandible: case report

C.Daoudi (1), N.Karich (1), A.Haloui (1), A. Bennani (1)

(1): Pathology departement, Univercity hospital Mohammed VI, faculty of medecine and pharmacy, Mohamed 1 univversity, Oujda, Morocco.

INTRODUCTION & AIM

Desmoplastic fibroma is an extremely rare and locally aggressive benign tumor. It can affect all bones of the body but most frequently the mandible, followed by long bones (the femur, radius, and tibia) and the bones of the pelvis.

Objective and importance : A unique aspect of our study is the rare focus on desmoplastic fibroma in children. Furthermore, reports of this pathology are exceedingly rare.

METHOD

We studied a 3-year-old patient, with no notable pathological history, who presented with swelling in the cheek area that has been developing since birth without pain or inflammatory signs.
The radiological examination showed a multilocular lesion breaking the bony cortex and reaching the jugal soft tissues.
A biopsy of the mass was performed intraorally.

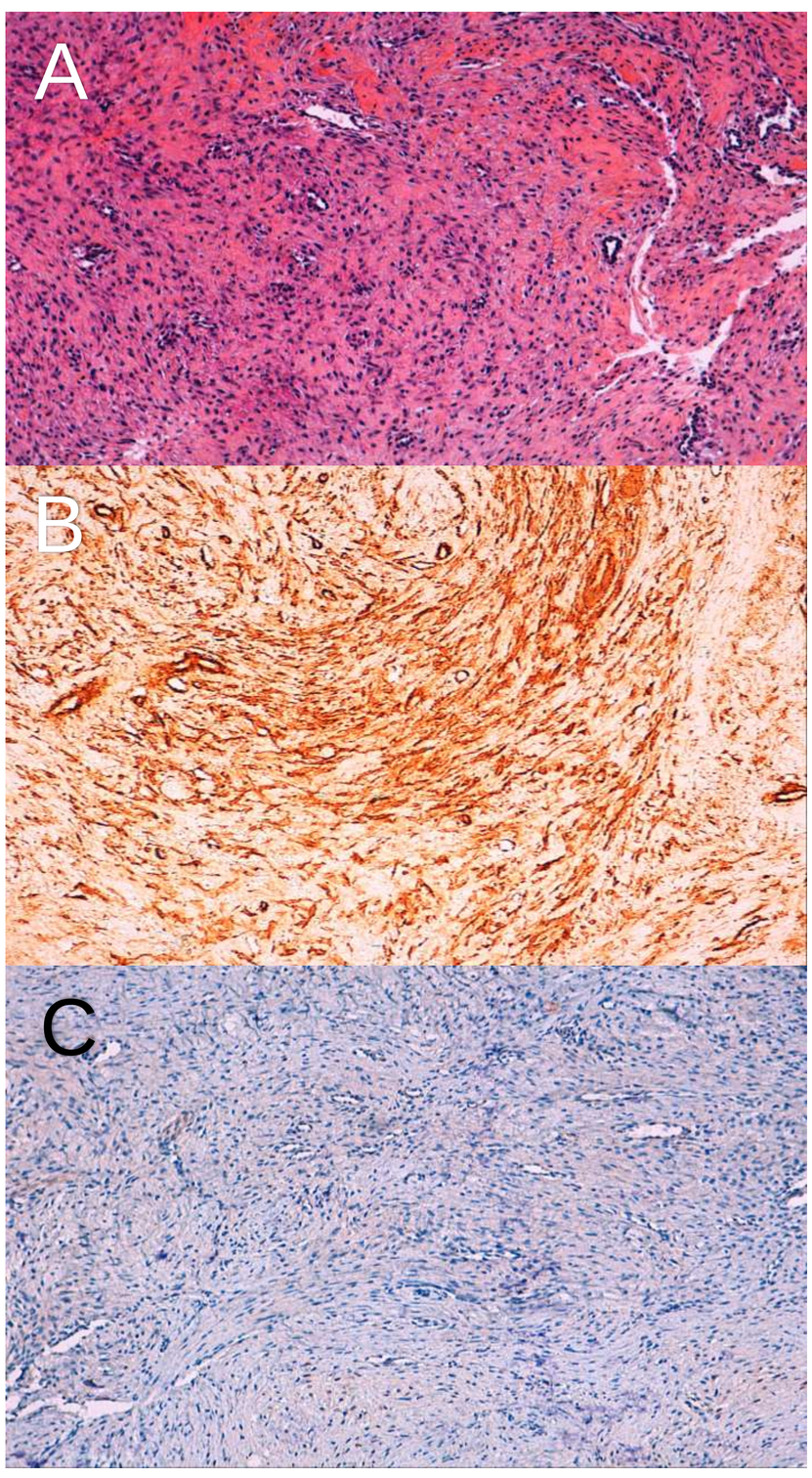

Figure: A) Histopathology section showing tumoral proliferation made of bundles of regular and uniform myofibroblastic cells with a collagenous stroma.
B) Positive marking of tumor cells by the anti-SMA antibody.
C) Negative marking of tumor cells by the anti-Desmin antibody.

RESULTS & DISCUSSION

Histologically, it is a tumoral proliferation made of bundles of regular and uniform myofibroblastic cells. The stroma is collagenous, the seat of a few congestive capillaries aligned parallel to the bundles. This proliferation is non-encapsulated and infiltrates the surrounding muscle and bone tissue.
The absence of cytonuclear atypia, atypical mitoses, and necrosis.
An immunohistochemical study was carried out showing:
- The positive marking of tumor cells using β-cathenin and AML.
By combining clinical, radiological, histological, and immunohistochemical data, the diagnosis of a desmoplastic fibroma was made.
Desmoplastic fibroma is an exceptional tumor that constitutes less than 0.1% of all primary bone tumors. It mainly affects adolescents and young people with a peak age between 20 and 30 years old and there is no gender predominance.
Clinically, desmoplastic fibroma is manifested by a painless swelling, difficulty in opening the mouth, and displacement of teeth. Sometimes, the tumor is asymptomatic and the discovery is incidental.
Radiologically, desmoplastic fibroma manifests as a well-defined, multilocular, and radiolucent lesion. Intralesional trabeculations are often present. Large lesions can destroy the bone cortex and infiltrate the surrounding soft tissue, which is clearly shown by MRI displaying a lesion with T1 and T2 hyposignal.
Histologically, it is a sparsely cellular-infiltrating tumor proliferation arranged in bundles of monomorphic spindle cells resting on the abundant collagenous stroma. It is characterized by the absence of necrosis and very rare mitoses. This proliferation may show positive cytoplasmic staining using β-cathenin and AML. The main differential diagnoses are fibrous dysplasia, low-grade central osteosarcoma, low-grade myofibrodysplasia, myoepithelial tumors, follicular dendritic tumors, and synovialosarcoma.
We selected surgical resection with wide resection margins as the treatment. Aggressive curettage may be used. Tumoral surgical margins were associated with higher recurrence rates.

CONCLUSION

Desmoplastic fibroma is an extremely rare, benign, and locally aggressive tumor that most frequently affects the mandible. It generally occurs in children and young adults in the form of an osteolytic mass. The positive diagnosis of histological and molecular biology is recommended to eliminate differential diagnoses. Treatment is based on surgery and the prognosis is characterized by recurrences in the case of excision with tumoral margins.

FUTURE WORK / REFERENCES

« Soft Tissue and Bone Tumours: WHO Classification of Tumours (Medicine): 5th edition »