Diagnosis and Management of Fibrous Dysplasia in the Pediatric Patient: Two Case Reports

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INTRODUCTION

Fibrous dysplasia (FD) is a benign disorder of bone in which areas of bone are replaced with a scar-like fibro-osseous tissue. The abnormal tissue can cause symptoms like bone pain, swelling, deformities, and fracture. FD can arise as a single lesion affecting one bone (monostotic) or as multiple lesions affecting many bones (polyostotic). On rare occasion, polyostotic FD may be present as a component of McCune-Albright syndrome, a multisystem genetic disorder characterized by polyostotic fibrous dysplasia, endocrine anomalies and café au lait pigmentation. Amongst all forms of FD, the craniofacial bones are among the most commonly affected locations. Diagnosis of FD is usually made during childhood when symptoms become apparent. However, presentation of the disease can vary and some cases of FD can go undiagnosed until adulthood.

CASE #1 – 10yo male

A 10 year old male with a past medical history of ADHD was referred to Cohen Children's Medical Center Department of Dental Medicine for a bony expansile lesion of the right maxilla, present for one year. The growth was described as gradual, painless, and not associated with any changes to the occlusion. Extraoral examination revealed a right midface growth with localized slight erythema. There were no other swellings present elsewhere on the body and café-au-lait spots were absent. Intraoral examination showed a maxillary right buccal cortical expansion with mild palatal expansion. Panoramic radiograph (Figure A) revealed a large, ground glass radio-opacity of the right maxilla involving the maxillary sinus and the right orbit. The dentition and mandibular condyles were in normal positioning. CT radiograph (Figure B) showed a well-defined expansile bony lesion with ground-glass radiodensity involving the right maxilla.

Under local anesthesia and nitrous oxide, an incisional biopsy of soft and hard tissue was obtained from the patient's right maxilla (Figure C) and sent for histopathological analysis. The final diagnosis was confirmed as benign fibro-osseous lesion compatible with Fibrous Dysplasia. As the patient was asymptomatic, neither surgical nor medical intervention was indicated and the patient elected for monitoring and regular follow-up visits.

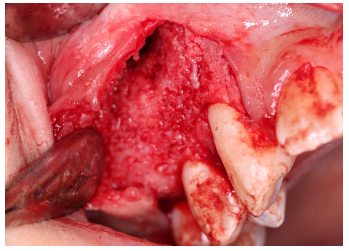

Figure C: A full thickness mucoperiosteal flap is opened to allow curetting of the bony lesion.

RADIOGRAPHS - CASE #1

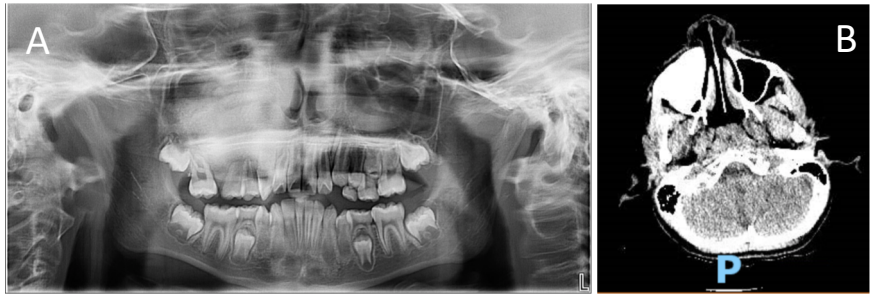

Figure A: Panoramic image. Ground-glass radio-opacity of the right maxilla
Figure B: CT scan coronal. Well-defined expansile bony lesion with ground-glass radiodensity involving the right maxilla.

RADIOGRAPHS - CASE #2

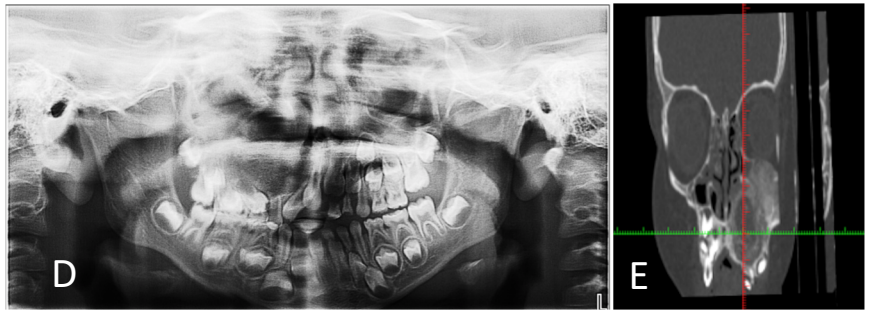

Figure D: Panoramic image. Ground-glass radio-opacity of the right maxilla
Figure E: CT axial view. Expanile ground glass lesion involving the right maxilla

CASE #2 – 5yo male

A 5 year old male with no significant past medical history presented to the Cohen Children's Medical Center Department of Dental Medicine for facial asymmetry on the right maxilla. He was experiencing nasal congestion and rhinorrhea but no other symptoms. Extraoral examination revealed a painless right midface fullness without overlying erythema. A 3x3.5cm café au lait spot was noted on the patient's lower chest. Intraoral examination showed a bony expansion in the area of the right buccal vestibule and right palate. Panoramic radiograph (Figure D) showed a well-defined ground-glass radio-opacity in the right maxillary sinus area. There was some displacement of the unerupted right-sided maxillary teeth. CT radiograph similarly showed an exansile ground-glass appearing lesion involving the right maxilla (Figure E). Under general anesthesia, an incisional biopsy sample was obtained from the patient's right maxilla and sent for histopathological analysis. Microscopic examination revealed several tissue fragments consisting of primitive pulpal tissue and follicular tissues (presumably of adjacent dentition), thinned cortical bone, and benign fibro-osseous lesional tissue. The fibro-ossesous component consisted of moderately cellular fibrous connective tissue composed of tapered spindle cells mixed with trabecula and circular bony elements. The bony component exhibited osteoblastic rimming which was interpreted as a function of the patient's age. The final diagnosis was confirmed as benign fibro-osseous lesion compatible with Fibrous Dysplasia. An endocrinology consultation was recommended for possible hormonal abnormalities associated with McCune-Albright syndrome. No surgical or medical intervention was indicated and the patient will return for regular follow-up visits.

DISCUSSION

The diagnosis of FD was established based on clinical, radiographic, and histopathologic features of the disease. For the asymptomatic pediatric patient, management largely involves regular follow-up and consultation with appropriate specialists. Medical and surgical intervention are to be considered if the patients become symptomatic, such as pain, pathologic fracture, or esthetic concerns. Management of FD is largely dependent on the clinical behavior of the lesions.

RESOURCES

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Lee, J S et al. "Clinical guidelines for the management of craniofacial fibrous dysplasia." *Orphanet journal of rare diseases* vol. 7 Suppl 1,Suppl 1 (2012): S2. doi:10.1186/1750-1172-7-S1-S2
Yang, Hsien-Yi et al. "Fibrous dysplasia of the anterior mandible: A rare case report." *Ci ji yi xue za zhi = Tzu-chi medical journal* vol. 30,3 (2018): 185-187. doi:10.4103/tcmj.tcmj_57_18