Bone reconstruction and optic functional restoration after fronto-orbital fibrous dysplasia resection
S Ferreira¹, LP Faverani¹, R Mendonça², FA Souza ¹, IR Garcia-Junior¹

Abstract

Introduction
Fibrous dysplasia (FD) is a non-neoplastic developmental hamartomatous disease of the bone in which fibrous tissue gradually expands and replaces normal bone. Clinical observations indicate that FD usually begins in childhood and chronically progresses throughout puberty and adolescence, and progression stops after adolescence in most cases. It can involve a single bone (monostotic FD), or multiple bones (polyostotic FD). Rarely FD may present with café-au-lait skin macules and endocrinopathies. This is known as the McCune-Albright syndrome. Once the diagnosis is established, the patient must be aware of the possibility of lesion manifestation elsewhere. The main treatment for craniofacial FD is surgery, which can be divided into conservative and radical resection.

Case Report
An 11-year-old female presented to our maxillofacial service with progressive disturbance of visual acuity and diplopia associated with left forehead prominence mouth with a one-year history of evolution. Computed tomography showed aspect of bone asymmetry with ground-glass appearance. Surgery for removal of her left forehead FD and decompression were performed. The immediate cranio-orbital reconstruction was performed using iliac and calvarium bone grafts and titanium mesh. Histopathological examination demonstrated proliferated fibrous formation in osteoid, which was compatible with fibrous dysplasia. An appropriate restoration of the facial morphology was clinically observed after one year. Her left visual acuity improved with a resolution of the diplopia.

Conclusion
In conclusion, we report a case of left forehead FD that showed slowly progressive visual symptom. Surgical treatment was effective in the preservation of vision and restoration facial aesthetics.

Introduction
Fibrous dysplasia (FD) is a non-neoplastic developmental hamartomatous disease of the bone in which fibrous tissue gradually expands and replaces normal bone including cranium. Although FD is a benign disease, it often results in severely craniofacial deformity, intracranial hypertension, or other kinds of dysfunction. If constriction of foramina or obliteration of bony cavities occurs, orbital dysopia, diplopia, proptosis, blindness, epiphora, strabismus, facial paralysis, loss of hearing, tinnitus, nasal obstruction, may also be evident. Histopathological appearance of the affected bone is reported to be composed by fibrous tissues that may contain focus of calcified bone, hyaline, cartilage, cysts and sometimes giant cells.

*Corresponding author
Email: sabrife@bol.com.br

¹ Division of Oral and Maxillofacial Surgery, Department of Surgery and General Clinic, Araçatuba Dental School, University Estadual Paulista Júlio de Mesquita Filho – UNESP Araçatuba, SP, Brazil
² Division of Neurosurgery, Santa Casa de Misericórdia Araçatuba, SP, Brazil

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Case report

Discussion
FD it is a rare benign bone disorder characterized by the replacement of normal bone for fibrous tissue intermixed with irregular woven bone. Usually manifests itself in the form of eyeball dislocation and visual disturbances when located near orbital cavity. Conservative bone contouring is most commonly used to improve facial aesthetics and prevent functional impairment. Conservative bone contouring, however, have a high recurrence rate ranging from 25% to 82% and surgery reintervention is often inevitable.

With the potential possibility of recurrence deciding the timing and extent of surgery is challenging. Although FD occurrence in cranio-facial location is relatively rare, ocular problems such as visual loss, diplopia and proptosis occur in 20–35% of patients. If these signs are detected, prophylactic surgical treatment is indicated before visual deterioration since acute visual disturbance is not reversible in these cases.

Therefore, the primary indications for surgery were restoration of ocular function, since visual symptoms progressed along the time; preservation of vision through decompression of the optic nerve and restoration of facial aesthetics. Although the chance of improvement of visual symptoms may be low when the optic canal are affected by FD for a certain period, our case indicated that it can be reversible by surgical decompression even one year after the initiation of the symptom.

Conclusion
We report a case of left forehead FD that showed slowly progressive visual symptom. Surgical treatment was effective in the preservation of vision and restoration facial aesthetics.

References
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