UPDATE

Craniofacial fibrous dysplasia surgery: A functional approach

E. Béquignon\textsuperscript{a}, C. Cardinne\textsuperscript{a}, X. Lachiver\textsuperscript{a}, I. Wagner\textsuperscript{a}, F. Chabolle\textsuperscript{a}, B. Baujat\textsuperscript{a, b, *}

\textsuperscript{a} Department of Oto-Rhino-Laryngology and Reconstructive Facial Surgery, Foch Hospital, 40, rue Worth, 92150 Suresnes, France
\textsuperscript{b} Department of Oto-Rhino-Laryngology and Reconstructive Facial Surgery, Hôpital Tenon, 4, rue de la Chine, 75020 Paris, France

KEYWORDS
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Summary

Background: Craniofacial fibrous dysplasia has not only esthetic but functional impact. Surgery is controversial, ranging from conservative to radical. It involves elevated hemorrhage risk, and should be progressive, based on an individual risk/benefit analysis with the aim of improving quality of life.

Case reports: Three patients (one male, two female; mean age, 35 years) with progressive orbital-temporal maxillary dysplasia were treated between 2008 and 2009 in our department. All showed exophthalmia and nasal obstruction. In one patient, symptomatology was aggravated by a frontal sinus cyst within the dysplasia. Another had associated auditory canal obstruction inducing recurrent external otitis. Optic nerve decompression was achieved on a combined coronal and endonasal approach, assisted by neuronavigation. Complementary remodelling resection, dacryocystorhinostomy and internal optic nerve decompression were performed. Functional results showed 70% improvement on a subjective scale for eye tension and nasal obstruction. Surgery was feasible in all patients, with no complications.

Conclusion: Current surgical management allies esthetic and functional concerns. Remodeling resection is the reference technique. The coronal approach is a good primary option for optic nerve decompression. Endonasal surgery with neuronavigation improves nasal ventilation and lacrimal canal permeability.

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Introduction

Fibrous dysplasia is a rare pathology, the first forms of which were described in 1872 and systematically presented by Lichtenstein and Jaffe \cite{1}. It is a benign condition, characterized by fibro-osseous proliferation of the facial bone and skull, similarly to ossifying or cemento-ossifying fibroma. Bone tissue is transformed into cellular fibrous tissue containing irregular trabeculae. It represents 3% of bone tumors and 7% of benign bone tumors. There are two forms: monostotic (70% of cases) and polyostotic (30%). Its physiopathology, described by Weinstein et al. \cite{2} in 2002, involves a genetic mutation on chromosome 20q13.
2.13.3, altering protein Gs. The mutation induces abnormal enlargement of cytokine IL6, involved in osteoclast differentiation. CT signs comprise bone hypertrophy straddling cranial sutures, with preserved bone concavity. Diagnosis is indicated by a juxtaposition of “frosted glass” condensation areas and areas of demineralization. On MRI, the lesion presents as variable isosignal on T1-weighted images, hyper-signal on T2 and heterogeneous gadolinium uptake. Onset is in childhood, with very slow evolution, causing asymmetric craniofacial deformity. It continues through the growth phase, and usually stabilizes after puberty, although sometimes continuing to evolve during adulthood. A few rare cases of malignancy have been described, at a rate of 0.5% to 4% depending on reports [3]. Fibrous dysplasia also causes functional complications: optic nerve compression with a risk of loss of vision, nasal and/or sinus obstruction, lacrimal tract obstruction, external auditory canal obstruction and impaired dental closure.

The present study sought to define treatment of the functional consequences of fibrous dysplasia, in the light of technical advances reported in the literature, illustrated by three clinical cases. The principle success criterion was improved quality of life.

Case reports

Patient 1

A 30 year-old man (Fig. 1) presented with right craniofacial fibrous dysplasia, evolving since the age of 5 years.

He had undergone several remodeling resection procedures involving the right maxillary, forehead and mandible, on oral or coronal approaches, since the age of 16 years. Functional complaints were moderate right nasal obstruction and discomfiting iterative external otitis and cerumen impaction. He had first been treated by external auditory canal reaming. One year later, he showed renewed right maxillomandibular growth inducing severe right nasal obstruction and ocular tension associated with right epiphora (Fig. 1). Maxillary remodeling resection was performed by neuronavigation-assisted degloving and dacryocystorhinostomy, with decompression of the internal orbital wall. Surgery improved overall symptomatology by 70% on a self-assessment scale, and exophthalmia was diminished.

Patient 2

A 40 year-old woman with right hemifacial fibrous dysplasia consulted for recent right-side headache associated with rapidly worsening exophthalmia following a series of major surgical interventions (Fig. 2). She had undergone several operations since the age of 15 years (mandibular osteotomy, then mandibulectomy associated to costal bone graft reconstruction) and, at the age of 36 years, right fronto-orbital remodeling with iliac bone graft. CT and MRI showed lysis within the dysplastic bone, suggestive of secondary right frontal cyst, with enhancement on gadolinium injection (Fig. 3).

Surgical revision was performed via a coronal approach with frontal skull bone flap. The liquid is evacuated from the cavity of a secondary cyst. A frontonasal canal was restored within the frontal dysplastic mass, and enlarged endonasally. The frontal access was closed by parietal grafts. Postoperative course was free of complications, with resolution of headache. Histology confirmed the secondary cyst. At 8 months, recurrent headache led to the discovery of obstruction of the frontonasal neo-canal; frontal sinus repermeabilization was achieved by neuronavigation-assisted endoscopy, which also confirmed the absence of cyst recurrence but found renewed fibrous dysplasia within the neo-cavity.
decompression via a coronal and right sub-tarsal approach, associated to bilateral canthopexy. Postoperative course was free of complications, with improved ocular comfort (Fig. 5). At 1 year, the nasal obstruction had worsened, to 80% discomfort; maxillary and piriform orifice remodeling resection was performed on a combined endonasal and endonasal approach (Fig. 5). Postoperative self-assessment showed a 70% improvement in nasal discomfort.

**Discussion**

The principles of fibrous dysplasia surgery are maximal tumor tissue resection, functional restoration, and esthetic optimization. The literature reveals five controversies concerning the management of functional complications in craniofacial dysplasia, summarized below with a view to drawing up an adapted treatment strategy.

**When to operate?**

In Kusano et al.,’s 2009 retrospective study of 11 patients treated from 1984 to 2006 [4], growth ceased in most lesions at adolescence, but in some cases, for no clearly identified reason, continued. Growth lasted longer in polyostotic (mean, 22.3 years) than monostotic (18.6 years) lesions. They drew the conclusion that it was necessary to wait for the end of the growth phase (i.e., adolescence) to perform remodeling in cases of monostotic involvement and that patients should be systematically warned of the risk of recurrence. There was no significant correlation between the bone location of the dysplasia and the duration of evolutivity. The present study confirmed that evolution may continue beyond adolescence. The mean age of the present three patients at treatment was 35 years. Our review of the literature suggested that surgery is indicated exclusively in case of symptomatic disease. Functional should take precedence over esthetic issues in indicating surgery [5]. The present three cases illustrate a progressive individualized strategy.

**Patient 3**

A 39-year-old woman with right maxillofacial fibrous dysplasia evolving since the age of 6 years had undergone remodeling resection at the ages of 11 and 21 years and presented under biphosphonates taken for 3 days every 6 months to treat recurrence of dysplasia. She consulted for symptomatic exophthalmia with right orbital pain (Fig. 4). Ophthalmologic assessment found conserved visual acuity with a convergence defect; she also showed moderate nasal obstruction, rated 30% on self-assessment (Fig. 4). A progressive strategy was adopted, giving priority to right orbital

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**Figure 2** Pre-operative photograph, patient 2: rapid evolution after exophthalmia.

**Figure 3** Pre-operative gadolinium-enhanced MRI, patient 2: lytic aspect with contrast uptake corresponding to secondary cyst; pre-operative CT scan, patient 2: cystic, lytic right frontal sinus aspect corresponding to secondary cyst.
adapted to the patient’s functional complaints. Onset of pain associated with rapid increase in volume suggests a simple or aneurismal cyst developing within the dysplasia. CT and MRI assessment should be performed. Treatment is surgical [6]. Case no 2 illustrates such specific cystic evolution, which needs to be recognized and treated as such. Rapid worsening of symptoms in parallel to progressive disease evolution is diagnostically suggestive.

What benefit is found with medical management?

Various medical treatments are recommended. Chapurlat (in 2006) administered a bisphosphonate injection (pamidronate 60 mg/day) for three successive days every 6 months for 18 months in case of pain and/or crisis [7], alleviating pain and reducing bone resorption in 50 % of cases. No mandibular osteonecrosis was observed. The author recommended stopping treatment 3 months ahead of scheduled surgery. In the present study, the patient did not seem to be stabilized under this regimen. Edgerton et al. and Ricalde and Horswell’s studies showed that progression was not definitely arrested [8,9]. Kruse et al., in a retrospective study of eight patients, recommended systematic administration of vitamin D (800 IU/day) and calcium supplementation (1 g/day) due to low serum levels [5]. Radiation therapy should not be applied in fibrous dysplasia, inducing a 400-fold elevation in malignancy risk according to Edgerton et al. (in 1985) [8].
Prophylactic versus therapeutic orbital decompression?

Surgical attitudes toward visual disorder in fibrous dysplasia are varied and controversial, notably as concerns prophylactic orbital decompression. Tan explained, in the Journal of Plastic and Reconstructive Surgery [10], the underlying mechanism, which consists in increased intraluminal pressure in the retinal vein due to external compression of the optic canal inducing arterial steal; also the optic nerve canal may be compressed by a cyst. Both mechanisms are found at work in the present series. Michael et al. [11], in a recent series, attributed loss of vision to a cause other than progressive optic canal compression. Lee et al., in a study published in the New England Journal of Medicine in 2002, reported that 95% of patients conserved normal vision despite optic canal stenosis [12]. Lee et al. and Michael et al., reviewing the literature, initially came out in favor of prophylactic decompression [11,12]. Tan, in a large retrospective study covering a 27-year period, published in 2007, compared prophylactic and therapeutic decompression [10] and, based on 18 patients, clearly concluded that regular visual assessment should be undertaken when stenosis is diagnosed; decompression is recommended only in case of continuous evolutive impairment of visual acuity. It arrests deterioration in more than half of cases, usually with actual improvement. Prophylactic decompression should not be performed in first intention, as impairment is found during the following year in a third of cases.

The contribution of advances in surgical technique?

Recently developed endonasal surgical material, including dedicated instrumentation with curved forceps, angulated reamers and neuronavigation, enhances surgical precision and technical possibilities. Advances in imaging, connecting a 3DMD surface-imaging camera up to a cone beam computed tomography (CBCT) system, allows evolution to be visualized by fusion of skin and bone images [13].

Conservative management or radical resection?

In 2009, Kruse et al. reported successful outcome in six out of eight patients managed by isolated remodeling resection [5]. This attitude is appropriate in evolutive craniofacial forms. The present study showed the interest of a progressive individualized approach in evolutive forms. The risk of recurrence being high, at 25%, functional impact should take precedence over esthetic considerations in guiding therapy.

Valentini et al., in 2009, reported a large series of 95 patients over the period 1980–2002 [14]. Twenty-two percent had polyostotic and 76% monostotic involvement. Sixty-one of the 68 patients managed by surgery had radical lesion resection; six received conservative treatment; one had 1-step mandibular resection with free peroneal flap reconstruction. There were no cases of recurrence following radical resection, and the author therefore recommended this radical but definitive solution in involuted monostotic forms. In contrast, Choi et al., in a 5-case study in 2009, demonstrated the interest of a non-invasive attitude, with a minimal vestibular or subciliary approach depending on the lesion, in involuted forms in which radical resection is precluded by skull-base involvement [13]. These findings taken together were drawn up into a decision tree for the management of functional complications (Fig. 6).

Conclusion

Craniofacial dysplasia requires progressive individualized multi-disciplinary management. The esthetic and above all functional consequences are to be treated surgically.
Remodeling resection is the reference technique. A coronal approach is suited to orbital decompression, while nasal and lacrimal duct permeability can be improved endonasally. Patients should be warned of the 25% recurrence risk. Active forms (about 20%) should be monitored on regular craniofacial CT.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References