ORIGINAL ARTICLE

Fibrous Dysplasia of Skull Base: Is There a Role of Endonasal Endoscopic Approach?

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Abstract

Fibrous dysplasia is abnormal proliferation of fibrous tissue interspersed with normal or immature bone. The records of 11 cases of fibrous dysplasia of paranasal sinuses with skull base involvement were reviewed. Six patients were polyostotic and 5 were classified as monostotic fibrous dysplasia. Four cases were managed endoscopically and only 1 patient had undergone right lateral craniotomy for relief of pressure symptoms. All the patients were regularly followed up to see for any recurrence. It was found that endoscopic clearance of disease in skull base should be reserved for patients with functional compression symptoms.

Keywords: Skull base tumor, endoscopic surgery, benign lesions of skull base.

INTRODUCTION

Fibrous dysplasia (FD) is a localized disorder of bone characterized by abnormal proliferation of fibrous tissue interspersed with normal or immature bone. Von Recklinghausen[1] in 1838 was first to give its pathologic description and Lichtenstein[2] suggested the term fibrous dysplasia in 1938 respectively.

About 1/4th cases of FD involve head and neck. Although FD involving temporal bone[3] has been reported widely but its impact on broader skull base with involvement on frontal, ethmoid and sphenoid sinuses is quite rare. Several external procedures have been used to manage these lesions but these days more conservative approaches are proposed. The management usually gets delayed until there are significant clinical symptoms or untolerable esthetic deformities. The aim of the present study was to compare the results of endonasal endoscopic approach in patients with FD of broader skull base along with external approaches.

MATERIAL AND METHODS

The records of 11 cases of FD of paranasal sinuses with involvement of skull base, treated in the department of Otolaryngology and Head and Neck surgery, Postgraduate Institute of Medical Education and Research, Chandigarh from January 2003 to December 2008 were reviewed. The information regarding clinical presentation, radiological investigation (Computerized tomography scan), treatment course and postoperative results was gathered and analyzed.

RESULTS

Out of 11 patients, 6 were polyostotic and 5 were classified as monostotic fibrous dysplasia. The 4 cases which were managed endoscopically had monostotic variant of FD. No patient had McCune Albright syndrome.

CLINICAL FEATURES

8 cases (72.7%) had vague symptoms like headache (ipsilateral in 6 and bilateral in 2), facial pain and broadened forehead. Nasal obstruction was present in 4 patients (36.3%) and 4 patients had proptosis (36.3%). Two cases had double vision and one had bilateral protrusion of eyeball (12.5%). Unilateral nasal obstruction was present in 3 patients (37.5%). Telecanthus was seen bilaterally in one patient and unilateral in 2 patients (37.5%) (Table 1).

ANATOMIC LOCATION OF THE LESION (RADIOLOGICAL)

The most commonly involved area of the skull base was ethmoid bone (75%). CT scan showed temporal bone thickening in 2 cases, thickening of cortex of occipital bone

Table 1: Clinical presentation

	Age (years)	Sex	Clinical symptoms	CT scan	Surgery	Postoperative	Complication	FU (years)
1	10	M	Proptosis, telecanthus (L)	Ethmoids, orbit (L) sphenoid B/L with skull base involvement	External ethmoidectomy following endoscopic clearance and optic nerve decopression	Proptosis absent	Revision endoscopic surgery	5.2
2	18	M	Nasal obstruction (L), proptosis	Frontal sinus skull base ethmoids B/L sphenoid	Endoscopic debridement GA	Nasal obst. headache, Proptosis reduced		3.5
3	17	M	Nasal obst, telecanthus and proptosis B/L	Frontal sinus unilateral (R) ethmoids B/L and skull base	Endoscopic debridement GA	Nasal obst. headache telecanthus absent	Revision endoscopic debridement	2.8
4	19	M	Nasal obst, proptosis	Sphenoid®, ethmoids (L), frontal sinus with skull base, (L) maxilla	Endoscopic debridement GA	Proptosis Headache absent		1.5
5	28	M	Vague headache	Temporal bone thickening (L)	Conservative treatment and observation	Headache reduced		5
6	30	F	Facial deformity and (L) facial pain	Broadened maxillary wall (L)	Observation	Facial deformity persisted but stable		3.2
7	19	M	No symptoms, diagnosed incidently	Thickened occipital bone	Observation			1.0
8	22	F	Mild nasal obst	Thickened anterolateral and medial wall maxilla (L)	Observation			0.8
9	12	M	Broadened forehead	Frontal bones thickened B/L	Observation			4.5
10	18	F	Vague hemifacial pain	(L) maxillary sinus walls thickened with hypertrophic mucosa	Conservative treatment and observation	Hemifacial pain absent		3.4
11	32	M	Ipsilateral headache	Temporal bone thickened (R)	Lateral craniotomy and subtotal resection	Headache off and on		2.5

in one, involvement of ethmoids with skull base in 4 (Figs 1 to 3), anterolateral wall of bilateral maxilla in two (Fig. 4), frontal bone involvement in one, frontal sinus with skull base involvement in four and sphenoid sinus involvement in 3 cases (bilateral in 2 and unilateral in 1).

SURGICAL OUTCOME AND FOLLOW-UP

Out of 11 patients, four were managed surgically by endonasal endoscopic approach, only 1 patient (case no. 11) had undergone right lateral craniotomy for relief of pressure symptoms and in one patient debridement was done using external ethmoidectomy approach. Rest 5 cases were managed conservatively.

All the patients were followed-up for a period ranging from 18 to 54 months with a mean follow-up of 3.3 years. All patients underwent CT scan six weeks after the surgery (Figs 1A and 2A).[3] (27.3%) patients had recurrence. One patient (case no. 1) who had undergone debridement by an

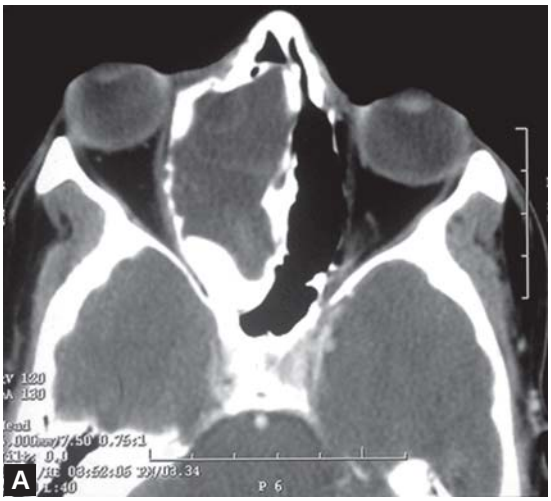

Figure 1A: Preoperative picture showing the ground glass appearance of ethmoids with skull base involvement

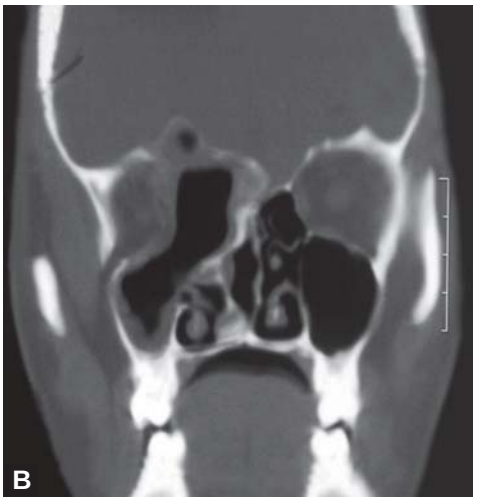

Figure 1B: Postoperative picture showing complete clearance of disease

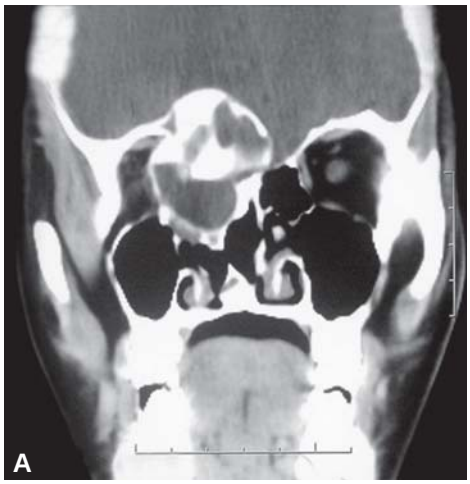

Figure 2A: Preoperative picture showing heterogenous soft tissue density with calcifications in posterior ethmoids

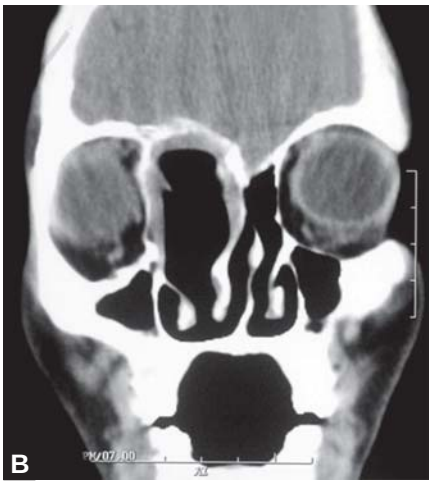

Figure 2B: Postoperative picture showing complete clearance of disease

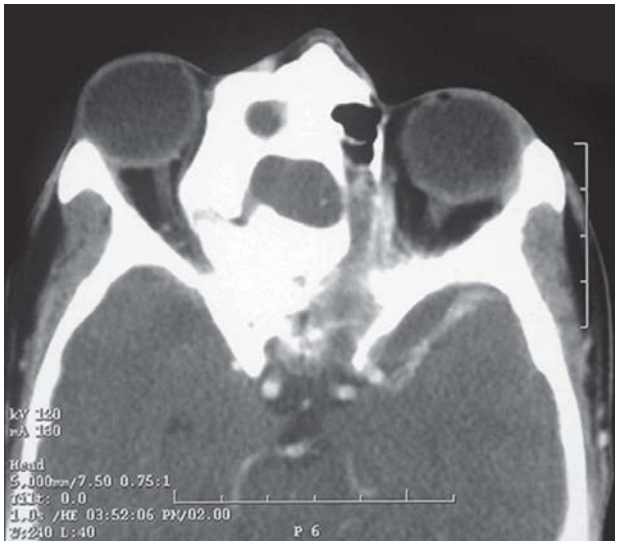

Figure 3: CT picture of fibrous dysplasia of ethmoids with intraorbital extension

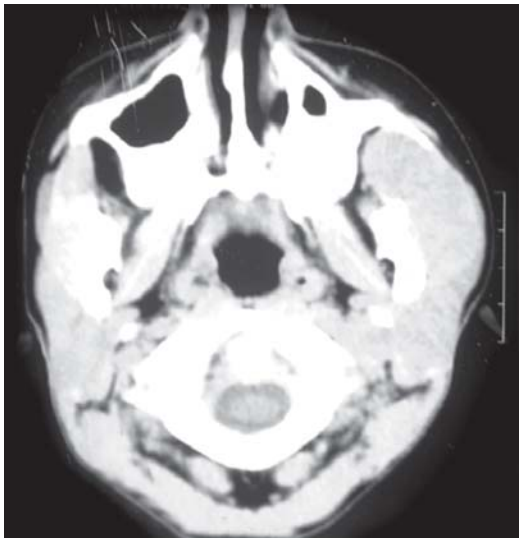

Figure 4: CT picture of fibrous dysplasia involving bilateral maxillae

external ethmoidectomy earlier, had recurrence of symptoms of proptosis and diminuition of vision after 1 year of follow-up for which a revision endoscopic debridement and optic nerve decompression was done. Another case (case no. 3) had recurrence of symptoms on left side and had undergone revision endoscopic debridement. Third patient (case no. 8) who underwent subtotal resection of the disease in maxilla on left side, had recurrence for which revision surgery was done. Only one patient (case 8) had facial deformity.

DISCUSSION

Out of 11 cases, we have taken into consideration those cases having involvement of the nose and paranasal sinuses along with skull base and who had undergone endoscopic resection of the disease. All were monostotic forms. Onset is typical in adolescence. The overall mean age was 20.4 years whereas the mean age at presentation was 15 years amongst 4 cases (case no. 1, 2, 5 and 8). Lawrence et al[3] reported the mean age of patients, 22 years and monostotic and polyostotic forms seen with equal frequency in males and females and high number of skull base involvement is seen with polyostotic FD. In our study, all 4 cases had monostotic FD and all were males. Spjut and coworkers[4] also noted more males than females. Lawrence et al[3] reported 6 patients with monostotic form and 15 with polyostotic FD.

The monostotic form, the mildest and most common form (70%),is noticed for lesions involving ribs and craniofacial bones and diagnosed between ages 20 and 30 years. The incidence of monostotic form may be even higher as they often remain asymptomatic. The polyostotic form (30%) has an early onset, seen in childhood, having more skeletal and craniofacial involvement. McCune Albright syndrome (3%) is the most severe form.

Van Tillburg[5] analyzed skull lesions from 144 patients in which frontal bones were more commonly affected followed by sphenoid, ethmoid, parietal and occipital bones. Another study[3] reported ethmoids as the most commonly involved bone followed by the sphenoid, frontal, maxilla, temporal, parietal and occipital bones in accordance with our study which also showed most common involvement of ethmoids with skull base radiologically.

In our study, pressure symptoms and displacement of orbital contents were seen in all the 4 younger age group patients which relates to the preponderance of ethmoids and sphenoid bone involvement.

Computed tomography is the study of choice for diagnosis and follow-up than plain X-rays as it gives superior and accurate extent of the lesion, differentiates from other osteodystrophies of the skull base. "Ground glass" appearance, symmetry, involvement of paranasal sinuses and thickness of the cranial cortices, involvement of sphenoid bone and orbital involvement can be seen with this modality. There is a clear margin between affected and unaffected bone.

In our study, all the four patients had undergone endonasal endoscopic clearance and debridement which was found adequate to control pressure symptoms. In 1st and the 8th patient, endoscopic subtotal resection was performed to relieve pressure symptoms only and facial deformity to be dealt later on as all patients were of pediatric and younger age group. All patients were watched conservatively, clinically and radiologically and revision surgery was performed once the symptoms recurred.

The other study[3] including 21 patients, 5 had undergone surgery to perform relief from proptosis either by external ethmoidectoy, lateral rhinotomy, facial degloving and craniofacial resection for extensive orbital rim involvement but no endoscopic resection was done. Four out of 5 had improvement in proptosis whereas our study showed disappearance of proptosis in 3 and reduction in 1 patient.

In other studies, temporal, occipital and parietal bone involvement has been reported[6,7] along with involvement of nose and paranasal sinuses but none in our 4 patients.

Fibrous dysplasia involving nose and paranasal sinuses with skull base involvement can present in myriad ways. Modern imaging modalities and histopathological analysis have made the diagnosis straight forward. Endoscopic clearance of the disease especially in the challenging areas like the roof and lateral wall of sphenoid and ethmoids with wide skull base involvement should be reserved in younger patients and for patients with functional and compression symptoms.

CONCLUSION

FD with skullbase is a benign condition. The surgery should be conservative with the primary goal being preservation of existing function.

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