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

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ORIGINAL ARTICLE

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 polystotic 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 in 1838 was first to give its pathologic description and Lichtenstein suggested the term fibrous dysplasia in 1938 respectively.

About 1/4th cases of FD involve head and neck. Although FD involving temporal bone 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 polystotic 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.

ANATOMIC LOCATION OF THE LESION (RADIOLOGICAL)
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).
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

Table 1: Clinical presentation

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

**CONCLUSION**

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

**REFERENCES**