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CASE REPORT
Craniofacial/Pediatric

Isolated Fibrous Dysplasia of the Bilateral Nasal Bones: Complex Management of the Bony Vault

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Summary: Fibrous dysplasia is a benign fibro-osseous process affecting the skeletal system, with resulting cystic and fibrous tissue expansion. Craniofacial fibrous dysplasia represents a small subset of monostotic disease, accounting for approximately 10%–25% of all such cases. Involvement of the frontal, temporal, and sphenoid bones has most commonly been described, with a limited number of reported cases citing disease isolated to the nasal bones. The case reported here is differentiated by the degree of expansion of the bilateral nasal bones and the required clinical management of the bony vault in the setting of gross nasal asymmetry.
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Fibrous dysplasia (FD) is an uncommon process of the skeletal system in which the bone is replaced by fibro-osseous tissue. This is radiographically represented by cortical thinning and a ground glass appearance on computed tomography (CT) imaging.[1,2] FD can result in loss of structural integrity, deformity, pain, and functional impairment.[3]

FD can be categorized into monostotic and polyostotic disease, affecting a single or several sites, respectively. Craniofacial involvement is frequently noted in polyostotic disease (50%–90%),[3] whereas it is less commonly observed in monostotic cases. Of the 10%–25% of monostotic cases affecting the craniofacial skeleton, most involve the maxilla, mandible, and bones of the cranium.[1,4]

FD can have a variable clinical course. It often presents before skeletal maturity, arising around 10 years of age.[3] Craniofacial FD has been found to develop particularly early, often citing lesion formation before the age of three.[3] Additionally, craniofacial FD differs from axial-based disease, in that lesions are slower growing. This often results in gradual, painless swelling and progressive asymmetry.[3] Other symptoms may include headache, facial pain, and auditory or visual impairment due to neurovascular impingement.[1]

Monostotic FD involving the nasal bones has only been described in two identified case reports, both demonstrating discrete lesions of the bony pyramid. We present a case with diffuse involvement of the nasal bones and discuss the management of the bony pyramid in the context of open septorhinoplasty for improved symmetry and function.

CASE REPORT

A 22-year-old man presented with significant nasal deformity (Fig. 1). He reported no history of nasal trauma and noted progressive asymmetry that had started in early childhood. Examination demonstrated nasal asymmetry, with a midline lesion affecting the entire bony pyramid. The area was nontender, solid in nature, and did not trans-illuminate on examination. Significant airflow obstruction was observed, left greater than right, with internal examination demonstrating severe leftward deviation of the septum. There was no evidence of external valve collapse, and bilateral Cottle maneuver elicited no improvement. CT imaging demonstrated overgrowth of the nasal bones; 3D rendering shows the extent of involvement of the nasal bones, with resulting rightward nasal deviation. (**See figure, Supplemental Digital Content 1**, which shows AP 3D reconstruction demonstrating bony overgrowth of the b/l nasal bones with resulting hypertrophy of the bony pyramid. **http://links.lww.com/PRSGO/D169**.)

SURGICAL CORRECTION

Open septorhinoplasty was planned to address nasal asymmetry, bulbous/under-projected tip, and polybeak

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Fig. 1. Preoperative assessment. A, Nasal analysis demonstrates a 22-year-old male patient with wide, asymmetric bony vault with bony overgrowth, widened mid-vault with asymmetric dorsal aesthetic lines and significant tip deviation. B, Lateral view demonstrates full radix, inadequate tip projection and a primary polybeak deformity. Photographic consent was preoperatively obtained from the patient.

deformity. While the surgical details regarding septal reconstruction, tip refinement, and alar flare correction are beyond the scope of this article, Supplemental Digital Content 2 illustrates the comprehensive plan used. [**See figure, Supplemental Digital Content 2,** which shows a Gunter graph demonstrating operative plan; asymmetric bony reduction (left > right), percutaneous osteotomies (low to low, oblique, and transverse radix), **http://links.lww.com/PRSGO/D170**.] While treatment of the bony vault is a standard aspect in many rhinoplasties, the degree of bony overgrowth and severe asymmetry required a complex strategy for correction. Dorsal rasping and medial/lateral osteotomies are often used for correction of dorsal convexity, nasal bone asymmetry, and excess nasal width.[5] However, in the senior author's assessment, these maneuvers were insufficient to allow for adequate correction in this case. Rather, in addition to the aforementioned maneuvers, oblique and transverse radix osteotomies were performed to allow for translocation of the entire bony pyramid in an effort to sync the axis deviation between the bony vault and the mid-vault/tip complex. When combined with medial and lateral osteomities, bony vault narrowing and translocation were achieved to improve overall symmetry. Finally, a bur was used (TPS, Stryker, Mich.) for further bony reduction and contour refinement.

POSTOPERATIVE FOLLOW-UP

The patient was evaluated 3-months postoperatively, where he was noted to have improvement in tip projection and correction of his primary polybeak (Fig. 2). Additionally, he demonstrated improvement in dorsal aesthetic lines, albeit with persisting asymmetry with fullness of the bony pyramid. The patient was noted to have significantly improved airflow postoperatively.

DISCUSSION

FD is an uncommon, benign process characterized by the replacement of mature marrow with abnormal fibro-osseous matrix.[2] It has a variable degree of craniofacial involvement, with prevalence differing significantly between monostotic and polyostotic disease. When it occurs, craniofacial FD typically involves the maxilla, zygoma, and cranium. To date, there is little information regarding cases affecting the isolated nasal bones.

When encountering mass lesions of the bony pyramid, surgeons should consider osteoblastoma, chondroblastoma, chondroma, ossifying fibroma, Paget disease, and fibrous dysplasia.[6] Although tissue biopsy is the gold standard for the diagnosis of any mass lesion, radiographic imaging can be used to demonstrate characteristics that are suggestive of FD. In patients who are asymptomatic or who have incidental lesions discovered, radiographic diagnosis is usually sufficient.[2] Further, CT imaging can be used to assess the disease stability. Maturing lesions demonstrate a radiolucent/radiopaque pattern, whereas stable/mature lesions demonstrate a ground glass appearance with diffuse opacities.[1,2]

When surgical intervention is being considered, the stability of the disease is paramount to long-term success. Monostotic disease is typically active until skeletal

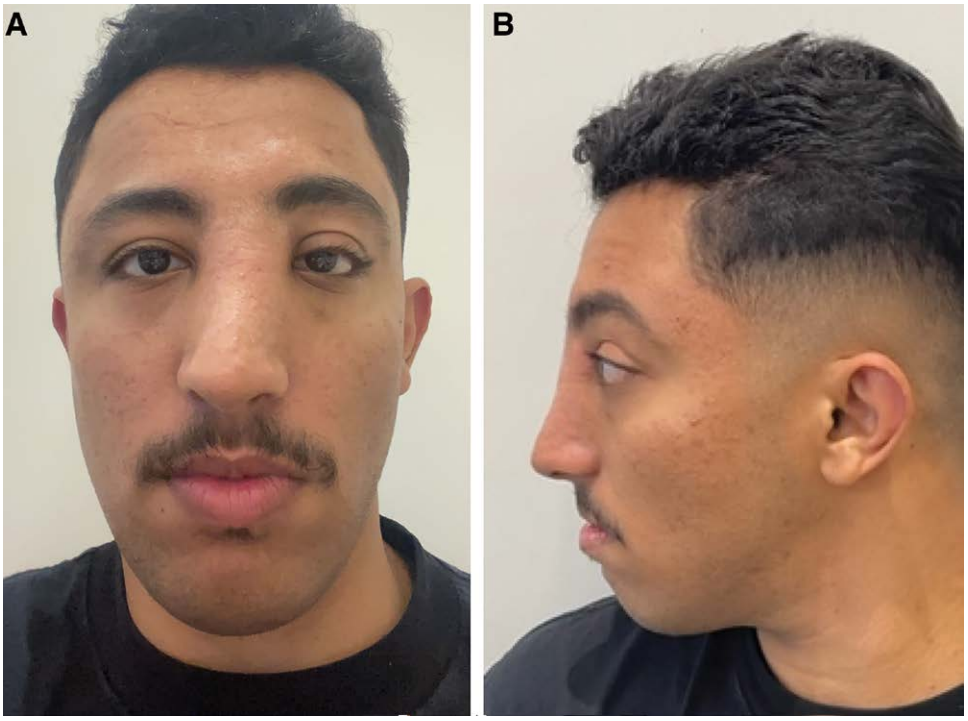

Fig. 2. Postoperative assessment. A, AP view demonstrating improved symmetry of the dorsal aesthetic lines with medialization of the bony vault and improved contouring of the hypertrophied nasal bones. B, Lateral view demonstrates correction of primary polybeak deformity with improved tip projection.

maturity, whereas polyostotic disease may progress further during adulthood.[2] As such, evaluating case characteristics and radiographic findings can help guide the decision to pursue, and timing of, surgery, as well as the specific operation indicated. In considering the role of surgery in craniofacial FD, Chen et al[7] advocated for a treatment algorithm based on anatomic zones, with the nasal bones included in zone 1. For lesions involving the naso-ethmoid complex, treatment is aimed at reducing airway obstruction and improving contour. This is often possible with subtotal excision, rather than radical resection, given the complex 3D nature of this region.[7]

In the case presented, subtotal reduction was pursued with these concepts in mind. This patient had long-standing stability of the lesion. Additionally, total lesion excision would have resulted in obliteration of the bony pyramid, necessitating complex reconstruction via open coronal approach with cantilever bone grafting. Further, this would not have addressed the patient's inadequate tip projection or polybeak deformity, both of which he found bothersome.

CONCLUSIONS

The case presented here represents just the third reported case of isolated FD of the nasal bones, with more extensive bony involvement. Considerations in the diagnosis and management include radiographic evaluation to assess disease stability, as well as to guide a treatment plan. An individualized treatment strategy should be devised based on disease stability, extent of involvement of surrounding structures, the sequelae of associated symptoms, and patient preferences.

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DISCLOSURES

Dr. Rohrich is the owner of Medical Seminars of Texas. He is also a consultant at Allergan/AbVie, Musculoskeletal Transplant Foundation, Galderma, InMode, and Evolus, and offers research support for Galderma, InMode, Merz, Cytrellis, Rion, and Teoxane. He receives book royalties from Thieme Publishers and instrument royalties from Eriem Surgical. All of the other authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

The patient provided written consent for the use of his image.

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