ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Professor of Clinical Neurosurgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Unique Case Observations

Editor
S. Ather Enam, MD, PhD
Aga Khan University, Karachi, Sindh, Pakistan

SNI
Open Access

Image Report

Hump on head: Rare case of bi-frontal fibrous dysplasia

Kunal Singla[1], Nishant Goyal[1], Pulkit Mittal[1], Zahed Ali Qamer[2], Jitender Chaturvedi[1], Arvind Kumar[3]

[1]Departments of Neurosurgery, [2]Pathology, [3]Pathology and Laboratory Medicine, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India.

E-mail: Kunal Singla - shinesingla47@gmail.com; *Nishant Goyal - drnishantgoyal@gmail.com; Pulkit Mittal - mittalpulkit.pm@gmail.com; Zahed Ali Qamer - thezaq27@gmail.com; Jitender Chaturvedi - drjittu28@gmail.com; Arvind Kumar - drarvindkumar10@gmail.com

***Corresponding author:**
Nishant Goyal,
Department of Neurosurgery,
All India Institute of
Medical Sciences, Rishikesh,
Uttarakhand, India.

drnishantgoyal@gmail.com

Received: 02 March 2023
Accepted: 14 November 2023
Published: 05 January 2024

DOI
10.25259/SNI_201_2023

Quick Response Code:

ABSTRACT

Background: FD is relatively rare in the craniofacial region, accounting for only 20% of all cases. Currently, two general subtypes of FD are recognized: monostotic and polyostotic. The monostotic form is more frequent, accounting for 75% to 80% of fibrous dysplasia cases.

Case Description: An 18-year-old male presented with the complaint of bony-hard swelling over the forehead for 8 years. Radiology showed an expansile osseous lesion involving frontal bones. The patient underwent bi-frontal craniectomy with gross total resection of tumour mass with titanium mesh cranioplasty. His postoperative period was uneventful and was discharged on the seventh postoperative day.

Conclusion: The cases of monostotic skull fibrous dysplasia should be treated by resection of the affected bone and cranioplasty. However, a more conservative re-contouring may be carried out in cases with multifocal involvement or when the excision is considered risky due to proximity to the major venous sinuses.

Keywords: Fibrous dysplasia, Frontal, Hump on the head, Parietal

INTRODUCTION

Fibrous dysplasia (FD) is a benign, congenital, and noncommunicable disease affecting both genders equally. A benign proliferation of fibrous tissue in the bone marrow characterizes it. FD is relatively rare in the craniofacial region, and incidence has been estimated at 1 in 5,000–10,000.[3]

The authors wish to report a rare case of monostotic FD, as it involves the bifrontal bone, overlying the superior sagittal sinus. We used the "cut, remove, and replace" method of dealing with FD and would like to discuss the management options of this rare entity.

CASE DESCRIPTION

An 18-year-old male presented with the complaint of a bony, hard swelling over the forehead for eight years. As per the patient's description, the swelling was "initially the size of a pea," but it gradually increased to "the size of a tennis ball." There was no history of rupture or discharge from it. This was associated with mild headaches without any aggravating or relieving factors. There was no history of seizure, vomiting, trauma, or blurring of vision. On examination, a bony, hard, nonpulsatile, immobile, non-tender swelling, 8 × 10 × 15 cm in size, was present over the midline, bifrontal region [Figure 1]. The rest of his neurological examination was unremarkable.

Computed tomography (CT) of the head showed an expansile osseous lesion involving the bifrontal bones with linear trabecular striations [Figure 2]. Magnetic resonance imaging (MRI) of the brain

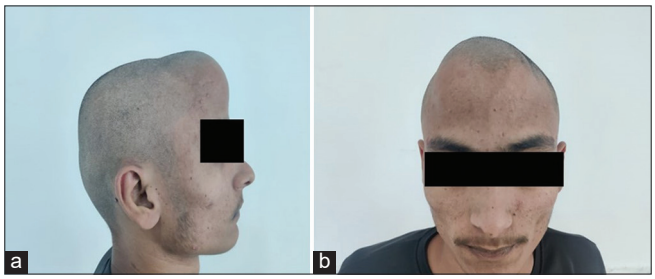

Figure 1: (a) Clinical photograph of patient lateral view and (b) front view.

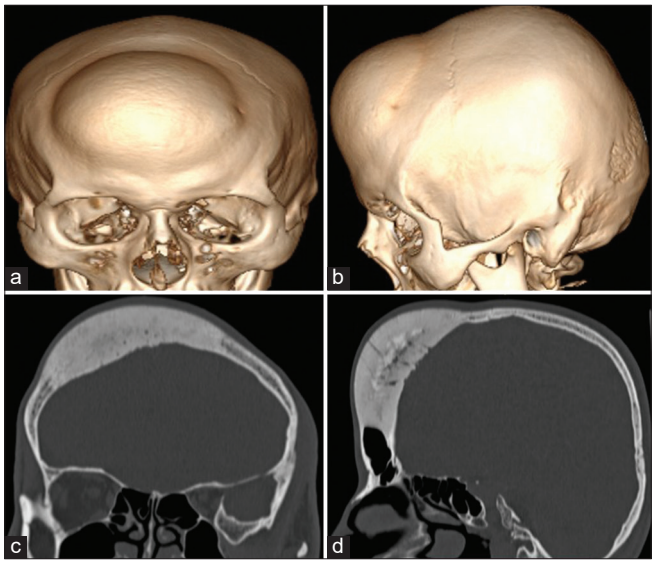

Figure 2: (a and b) Three-dimensional computed tomography (CT) scan reconstruction showing frontal bone hump. (c and d) Bone window of coronal and sagittal CT scan.

revealed a swelling in the frontal region that was seen involving both the inner and outer table of the frontal bone, causing the widening of the diploic spaces [Figure 3]. No significant enhancement was seen on postcontrast images.

The tumor was resected by bifrontal craniotomy [Figure 4a]. Burr holes were made circumferentially around the lesion, avoiding the midline and leaving some margin. Gradually, the dura was separated circumferentially, avoiding any sinus injury. The tumor mass was enveloped in the bone (both inner and outer tables). Following this, cranioplasty was done using titanium mesh (Johnson and Johnson, India), fixed with miniplate and screw [Figure 4b].

The postoperative period was uneventful, and the patient was discharged on the 7th postoperative day. Histopathology revealed a fibro-osseous lesion with irregular curvilinear woven bone without conspicuous osteoblastic rimming, intervening fibrous tissue composed of bland fibroblastic spindle cells [Figure 5]. It was suggestive of benign fibro-osseous lesion favoring FD. He is doing well at a 12-month follow-up that shows an excellent cosmetic result [Figure 6] without any recurrence of symptoms.

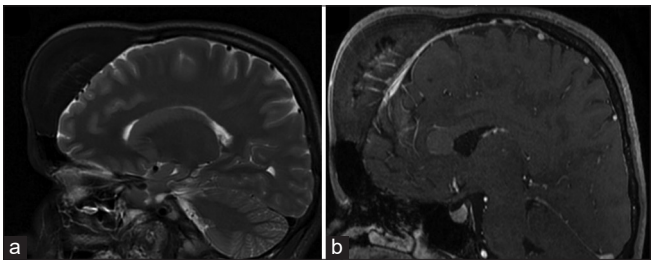

Figure 3: (a) Magnetic resonance imaging T2 sagittal and (b) T1 contrast sagittal.

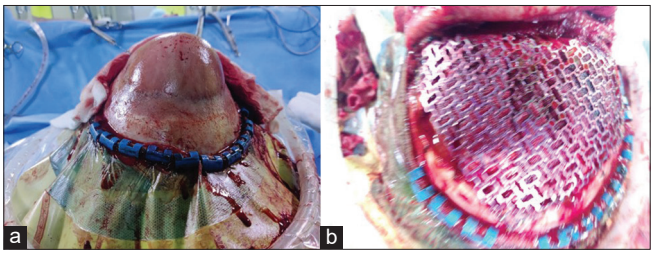

Figure 4: (a) Intraoperative image after skin flap elevation, (b) after resection and placement of mesh over the craniectomy defect.

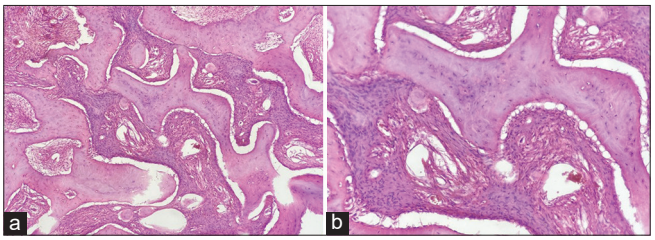

Figure 5: (a and b) Fibro-osseous lesion with irregular curvilinear woven bone without conspicuous osteoblastic rimming, intervening fibrous tissue is composed of bland fibroblastic spindle cells (H&E).

DISCUSSION

FD is characterized by fibro-osseous lesions with blending of fibrous and osseous tissue, with resultant secondary bony metaplasia, without osteoblast maturation producing immature, newly formed, and weakly calcified bone.[7] It usually presents as a painless swelling, which progressively increases in size.

CT best characterizes FD. Its appearance pattern on CT may vary because of variable combinations of osseous as well as fibrous elements. It may be ground glass or mixed (56%), homogeneous radiopaque (sclerotic) (23%), and radiolucent with a lytic pattern of trabeculae (cystic) (21%).[2] On MRI, FD shows homogenous, moderately low-signal intensity on T1 and T2-weighted images. Most of the lesions display moderate-to-high central contrast enhancement with some rim enhancement. The degree of contrast enhancement on T1-weighted images depends on the amount and degree of bone trabeculae and collagen present. Likewise, in our case, there was no contrast enhancement.[1]

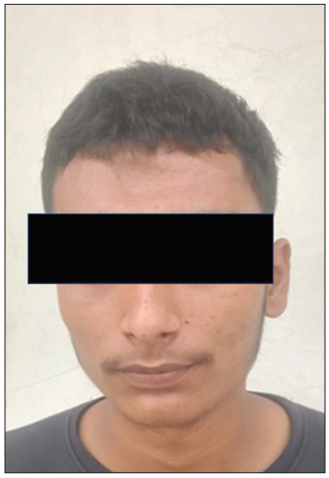

Figure 6: Clinical photograph of the patient at 12-month follow-up.

The aim of the treatment is correction of the facial deformity, prevention of pathological fracture, control of pain, and prevention of recurrence.[5] Radiation therapy and chemotherapy have no role in the treatment of this disease, and radiation therapy may increase the risk of malignant transformation.[4] Surgery is the mainstay of treatment in FD. We carried out a radical resection with mesh cranioplasty to correct the deformity to prevent recurrence.[6]

CONCLUSION

To conclude, cases of monostotic skull FD should be treated by resection of the affected bone and cranioplasty. As we have shown, even a case involving the bifrontal bones and lying over the superior sagittal sinus can be resected completely, followed by reconstruction. However, a more conservative recontouring may be carried out in cases with multifocal involvement or when the excision is considered risky due to proximity to the major venous sinuses.

Ethical approval

Not applicable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Chandavarkar V, Patil PM, Bhargava D, Mishra MN. A rare case report of craniofacial fibrous dysplasia. J Oral Maxillofac Pathol 2018;22:406-9.
2. Gupta D, Garg P, Mittal A. Computed tomography in craniofacial fibrous dysplasia: A case series with review of literature and classification update. Open Dent J 2017;11:384-403.
3. Javaid MK, Boyce A, Appelman-Dijkstra N, Ong J, Defabianis P, Offiah A, *et al*. Best practice management guidelines for fibrous dysplasia/McCune-Albright syndrome: A consensus statement from the FD/MAS international consortium. Orphanet J Rare Dis 2019;14:139.
4. Maher CO, Friedman JA, Meyer FB, Lynch JJ, Unni K, Raffel C. Surgical treatment of fibrous dysplasia of the skull in children. Pediatr Neurosurg 2002;37:87-92.
5. Menon S, Venkatswamy S, Ramu V, Banu K, Ehtaih S, Kashyap VM. Craniofacial fibrous dysplasia: Surgery and literature review. Ann Maxillofac Surg 2013;3:66-71.
6. Ozek C, Gundogan H, Bilkay U, Tokat C, Gurler T, Songur E. Craniomaxillofacial fibrous dysplasia. J Craniofac Surg 2002;13:382-9.
7. Xu J, Li X, Lv CS, Chen Y, Wang M, Liu JF, *et al*. Treatment protocols for growth hormonesecreting pituitary adenomas combined with craniofacial fibrous dysplasia: A case report of atypical McCuneAlbright syndrome. Exp Ther Med 2014;8:877-80.

How to cite this article: Singla K, Goyal N, Mittal P, Qamer ZA, Chaturvedi J, Kumar A. Hump on head: Rare case of bi-frontal fibrous dysplasia. Surg Neurol Int. 2024;15:1. doi: 10.25259/SNI_201_2023