Management strategies of fibrous dysplasia involving the paranasal sinus and the adjacent skull base

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April 05, 2024

Abstract

Objective: The current management of fibrous dysplasia (FD) involving the paranasal sinuses and the adjacent skull base is controversial. This study is to present our experience in the management strategy of FD involving the paranasal sinuses and the adjacent skull base. Design/setting: 23 patients from 2006 to 2019 with monostotic fibrous dysplasia (MFD), polyostotic fibrous dysplasia (PFD), or McCune-Albright Syndrome (MAS) involving the paranasal sinuses and the adjacent skull base were retrospectively reviewed. This study series was divided into 3 groups based on the management strategies: the observation group, the surgery group, and the optic nerve decompression group. Main outcome/results: The observation group included 9 asymptomatic MFD patients with a stable condition during the follow-up period of 15 to 164 months. The surgery group included 10 symptomatic MFD patients having personalized endoscopic endonasal surgery. The patients' symptoms were relieved after surgery. The optic nerve decompression group included 4 patients with visual loss, who underwent endonasal endoscopic optic nerve decompression with the aid of image-guided navigation. Their vision improved after surgery. Conclusion: The clinical observation and periodic computed tomography (CT) scan is adopted for asymptomatic patients. Surgery is indicated in symptomatic patients. Optic nerve decompression is recommended as soon as possible if the patient has visual loss, whereas prophylactic decompression is not recommended if the optic nerve is encroached upon by FD without visual loss. Navigation plays an important role in endoscopic surgery involving the paranasal sinuses and the adjacent skull base, especially in FD resection and optic nerve decompression.

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Keywords: fibrous dysplasia (FD), paranasal sinus, skull base, functional endoscopic sinus surgery (FESS), navigation-assisted surgery

Key Points :The clinical observation is adopted for asymptomatic patients. Surgery is indicated in symptomatic patients. Navigation plays an important role in endoscopic surgery in FD resection and optic nerve decompression.

Introduction

Fibrous dysplasia (FD) of bone, also referred to as fibrous hyperplasia of bone, is a group of non-neoplastic diseases with fibro-osseous bone lesions. FD is a developmental disorder induced by abnormal proliferation of fibroblasts resulting in replacement of normal cancellous bone by structurally immature osseous tissue[1-3]. FD may involve any bone of the body and affect one or more bones. It is divided into three types: monostotic FD (MFD), polyostotic FD (PFD), and McCune-Albright syndrome (MAS). When the lesion involves the craniofacial skeleton, the skull base is the most common site of involvement[1-3].

Because of the low incidence rate, diverse natural history and complicated surgical procedure, at present, the management of FD involving the paranasal sinuses and the adjacent skull base is controversial. Therefore, many scholars have called for development of a global consensus on improving patient management[4-7]. In this study, we showed our experience and treatment strategies for the management of 23 FD cases which involved the paranasal sinuses and the adjacent skull base.

Material and Methods

2.1 Subjects

This study included 23 patients with FD involved in at least one paranasal sinus bone and in the adjacent skull base. There were 19 cases of MFD, 3 cases of PFD, and 1 case of MAS. The 23 patients included 12 females and 11 males. The median age was 33 (range, 2-67) years. The common symptoms were nasal obstruction (13 cases, 56.6%), rhinorrhea (9 cases, 39.1%), headache (7 cases, 30.4%), facial pain (6 cases, 26.1%), visual impairment (4 cases, 17.4%), snoring (5 cases, 21.7%), facial deformity (3 cases, 13%), and obvious exophthalmos (2 cases, 8.7%). There was no obvious epistaxis or diplopia. The clinical characteristics of the patients were listed in Table 1, 2 and 3.

This study was approved by the Ethics Committee of our hospital and conducted with written informed consent from patient.

All the patients received a thin slice computed tomography (CT) scan of the paranasal sinuses, including coronal and axial views. The CT scan revealed the lesion location and was helpful for diagnosis. The most common findings of FD on CT are an expanded bone with ground-glass appearance. The bone window showed that the image of a normal bone's structure was replaced by the image of an ivory high-density pattern (Figure 1), a low-density pattern (Figure 2) or a frosted glass pattern (Figure 3A and 3B).

2.2 Management strategies

This study series was divided into 3 groups based on management strategies, namely, the observation group, the surgery group, and the optic nerve decompression group.

The observation group included 9 patients with asymptomatic MFD, and adopted the method of clinical observation and regular fellow-up. The patients received a paranasal sinus CT scan periodically. The clinical data were shown in Table 1. Three of the nine patients had chronic rhinosinusitis, who received functional

endoscopic sinus surgery (FESS) only for their chronic rhinosinusitis. The other six patients were found with FD on physical examination. Two of them received a biopsy through an endoscopic endonasal approach.

The surgery group included 10 patients with symptomatic MFD. The clinical data were shown in Table 2. Five of the patients underwent FESS for their chronic rhinosinusitis simultaneously. FD was relatively hard and was removed by an electric drill (Medtronic IPC, USA)and Kerrison Rongeurs. Seven of the cases were excised with the aid of image-guided navigation (Fusion, Medtronic ENT, USA) (Fig. 3C, 3D).

The optic nerve decompression group included 4 patients with vision loss, who underwent endonasal endoscopic optic nerve decompression with the aid of image-guided navigation. The clinical data were shown in Table 3. One of the patients received bilateral endoscopic optic nerve decompression simultaneously. The MAS patient accepted the first surgery of left endoscopic optic nerve decompression in February 2016 and again in July 2017 (Fig. 3A and 3B).

Result

The patients in the observation group were followed up regularly. For example, in case one in table 1 the paranasal sinus CT scan in June 2008 showed ivory high-density pattern sphenoid bone FD, then the patient received endonasal endoscopic biopsy and was diagnosed as FD by histopathological examination. He was asymptomatic and followed up regularly. The paranasal sinus CT taken in 2008 was compared with the paranasal sinus CT reviewed in 2016, which revealed that the FD was in a static state (Fig.1).

In the surgery group, five patients with ethmoid bone FD underwent total resection, while three patients with sphenoid bone FD underwent subtotal resection and two patients underwent partial resection through an endonasal endoscopic approach. Five of the patients underwent FESS for their chronic rhinosinusitis simultaneously. The symptoms of the patients were relieved after surgery, and there were no intracranial or intraorbital complications. The patients in this group were followed up for 1-13 years, and there was no recurrence.

During the operation, we found that although some MFD lesions were large, neither the germinal center nor the pedicle was large. A thin layer of fibrous tissue separated FD from normal bone, and it was relatively safe to dissect along this layer of fibrous tissue. For example, case three in Table 2 is a two-year-old boy. The patients' paranasal sinuses CT scan showed that the left ethmoid sinus and nasal cavity were filled with a large (40.5mm×6.3mm×29.9mm) and highly dense mass (Fig. 2). The mass originated from the left cribriform plate, compressed the left lamina papyracea, and extended to the bottom of the nasal cavity. The frontal sinuses and sphenoid sinuses were undeveloped. The thin layer of fibrous tissue was found between the FD and the partial cribriform plate and lamina papyracea during the surgery (Fig. 2). This phenomenon was also seen in the resection of sphenoid bone FD (Fig. 3C).

In addition, MFD may have degenerative changes with age, as shown in case 4 and case 5 (Fig. 3D) in table 2. The paranasal sinus CT of case 5 revealed frosted glass changes of the left sphenoid pterygoid process and great wing, with a 2.3× 1.0 cm soft tissue shadow. The pterygoid process and great wing change was diagnosed as FD and the 'soft tissue shadow' change was determined to be hyaline degeneration by pathological examination.

In the optic nerve decompression group, the effects of optic nerve decompression were shown in Table 3. The youngest patient in this group, a 10-year-old girl with MAS, showed left visual loss again one and half a years after her first left optic nerve decompression. The patient received left optic nerve decompression again, and the left visual acuity remained stable so far. However, the CT showed that the right optic nerve was encased by FD, but the patient's right eye vision remained stable all the time (Fig.3A and 3B). Case 1 in Table 3 underwent bilateral optic nerve decompression due to cystic degeneration from FD (Fig. 3E). The CT scan of 2 years before surgery revealed that the left optic nerve was completely encased and the right optic nerve was partially encased by FD (Fig. 3F, G, H), but the vision was normal until 1 month before the surgery.

Discussion

In view of the rarity, complex and varied natural history, and clinical manifestation of FD, it is difficult for clinicians to develop a standardized treatment plan. As a consequence, no treatment guidelines currently exist [1]. Although there was not an international guideline for the management of FD/MAS, an international workshop has been set up to focus on improving FD/MAS management and understanding the importance of a multidisciplinary team for the lifetime management of FD/MAS [7].

FD treatment includes clinical observation, medical therapy and surgery. Medical treatment for FD is limited to relieving a patient's symptoms. One of these medications is bisphosphonate, which may help to improve the function, relieve the pain, and reduce the fracture risk. However, a recent randomized, double blind, placebo-controlled trial disputed these effects [8,9]. Radiotherapy is contraindicated owing to high prevalence of malignant transformation. Therefore, clinical observation and surgery are currently the main treatment strategies for FD.

Clinical observation is adopted for asymptomatic FD patients, but the patients should be required to have periodic radiologic evaluations. As FD is a slow-growing lesion, tends to be stable after puberty, and has a low malignancy potential, most of the specialists recommend clinical observation if there are no symptoms, but periodic imaging should be performed to confirm that there is no progression or regrowth in follow-up [1]. Comparing craniofacial FD management between 1980 and 2002 with management between 2003 and 2013 showed that observation has replaced surgery as the most used method. Watching carefully and attentively was indicated in cases of stable lesions, and it was the best therapeutic option, if possible [4]. In France, with the establishment of National Reference Centers, 57 specific recommendations have been provided for the diagnosis, prognosis, and follow-up of patients with FD/MAS. If the skull and/or facial bones are involved, a skull CT is recommended to accurately evaluate the risk of neurological compromise due to alterations of the foramina. MRI should be viewed as a second-line imaging study. Radiographic monitoring is recommended every 2-3 years for follow-up patients [10]. It was demonstrated that the characteristics of FD on CT and the natural radiographic progression may vary from a "ground-glass" or homogenous appearance to a mixed radio-dense/radio-lucent lesion as the patient ages [2]. Based on preoperative radiology, the sensitivity and specificity to correctly detect FD were 54.6% and 96.9%, respectively.

Lesions with classic ground glass appearance or mixed pattern on CT should not warrant diagnostic biopsy, especially in asymptomatic patients. However, if radiologic diagnosis is uncertain, biopsy may be warranted to arrive at the definitive diagnosis based on radiographic and histologic correlation [11]. Biopsy of a lesion does not specifically induce growth of FD, but if the lesion is asymptomatic, and/or in the cranial base, a biopsy may not be necessary[2].

Surgery is indicated in symptomatic patients. Although application of surgical treatment for craniofacial FD is controversial, many publications have provided views on the surgical treatment of FD[12-14]. Comparing craniofacial FD management between 1980 and 2002 with management between 2003 and 2013 revealed that radical resection (if possible) of FD was the only technique to obtain resolution of the disease [4]. The advantages of surgical treatment of FD should be appropriately weighed against possible complications [1]. The aim of the surgical treatment for craniofacial FD is to remove the bulk of the lesion, reserve the cranial nerve compression, and resolve the aesthetic problem[6]. The extent of the resection should be based on the location of the pathological bone and its proximity to important structures, as radical or complete resection may not be necessary or possible [5]. Treatment protocols should be tailored to individual patient's needs, with the aim to achieve the best possible esthetic and functional outcome with the least postoperative morbidity[15-16]. Therefore, surgical treatment planning must take into account several factors (i.e., natural history of the disease, presence of symptoms, site of the lesion, and the relationship with critical anatomic structures) [1].

Early surgery is necessary for the patient with MFD when the lesion was limited to ethmoid or sphenoid bone with obvious symptoms. It is because the growth and expansion of the FD may lead to the obstruction of the sinus ostia and result in rhinosinusitis and mucocele. Furthermore, it has been reported that the lesion may oppress the adjacent bone, such as the lamina papyracea and the anterior skull base, extend into the orbit and intracranial cavity, and ultimately lead to epiphora, diplopia, proptosis, impairment of visual acuity, meningitis, cerebrospinal fluid leak, and so on [17]. Five patients with ethmoid bone FD

and three with sphenoid bone FD in the surgery group of this study series underwent total resection or subtotal resection through an endonasal endoscopic approach respectively. Only two patients with sphenoid bone FD underwent partial resection through an endonasal endoscopic approach. And five surgery group patients underwent FESS for their chronic rhinosinusitis simultaneously. Meanwhile, what we found in these procedures was that in the process of FD expansion toward the cavity of a paranasal sinus, the epithelial tissue in the sinus cavity may be compressed into a thin layer of fibrous tissue, which separated FD from normal bone, and it was relatively safe to dissect along this layer of fibrous tissue. Combined with use of intraoperative image-guided navigation and advanced surgical instruments, these surgeries were performed successfully without intracranial or intraorbital complications.

For FD patients with impaired vision, it was recommended to have optic nerve decompression as soon as possible. However, if FD was encroaching on the optic nerve without impaired vision, prophylactic decompression is not recommended. This is the current consensus[1,18-20]. A retrospective analysis in 91 patients with craniofacial FD involving the optic nerves showed that 17% of nerves were less than 50% encased, 22% were 50-99% encased, and 61% were 100% encased. Yet optic nerve decompressions were performed in only 13 patients (6 prophylactic and 7 therapeutic) since the authors regarded that the majority of optic nerves encased with FD did not present symptoms of optic neuropathy and appeared to be stable over time[18]. Satoh K, et al. held that current strategies should focus on esthetic improvement, with careful observation carried out to assess for optic canal encroachment without prophylactic decompression [19]. In another study, in asymptomatic patients, stable vision occurred in 76% of patients receiving decompression and 95% of patients not undergoing surgery ($P< 0.001$). Vision impairment may be associated with the concomitant presence of cystic lesions (i.e., mucocele, hemorrhage, and aneurysmal bone cyst) [20]. Holl DC, et al. reported a case of FD in which the patient developed an aneurysmal bone cyst leading to left optic nerve compression with an acute visual loss. An emergency optic nerve decompression resulted in complete restoration of vision. [21]. In this study, one patient had acute bilateral visual loss due to cystic degeneration formation. Another patient with MAS underwent left optic nerve decompression twice, but in fact, although the CT showed that the right optic nerve was encased by FD, the right eye vision of the patient remained stable all the time.

Navigation plays an important role in paranasal sinus and adjacent skull base FD resection, and in optic nerve decompression. Stereotactic navigation was recommended, as the FD/MAS process often distorted normal intranasal landmarks used in sinus surgery [5]. Navigation assisted, endonasal endoscopic optic nerve decompression was usually effective for the treatment of nontraumatic optic neuropathy[22].

Conclusion

Clinical observation is adopted for asymptomatic patients with FD, but the patients should be required to undergo periodic radiologic evaluations. Surgery is indicated in symptomatic patients. In FD patients with visual change or vision loss, an optic nerve decompression was recommended as soon as possible; however, if FD is encroaching on the optic nerve without impaired vision, prophylactic decompression is not recommended. Navigation plays an important role in paranasal sinus and adjacent skull base FD resection and optic nerve decompression.

REFERENCES

1. Schreiber A, Villaret AB, Maroldi R, Nicolai P. Fibrous dysplasia of the sinonasal tract and adjacent skull base. *Curr Opin Otolaryngol Head Neck* 2012;Surg 20:45-52.
2. Lee JS, FitzGibbon EJ, Chen YR,Kim HJ, Lustig LR, Akintoye SO, Collins MT, Kaban LB. Clinical guidelines for the management of craniofacial fibrous dysplasia. *Orphanet J Rare Dis* 2012;7:S2.
3. Amit M, Fliss DM, Gil Z. Fibrous dysplasia of the sphenoid and skull base. *Otolaryngol Clin North Am* 2011;44:891-902.
4. Valentini V, Cassoni A, Terenzi V, Della Monaca M, Fadda M T, Rajabtork Zadeh O, Raponi I, Anelli A, Iannetti G. Our experience in the surgical management of craniofacial fibrous dysplasia: what has changed in the last 10 years? *Acta Otorhinolaryngol Ital*2017;37:436-443.
5. DeKlotz TR, Kim HJ, Kelly M,Collins MT. Sinonasal disease in polyostotic fibrous dysplasia and

McCune-Albright Syndrome.*Laryngoscope* 2013;123:823-828.
6. Lei P, Bai H, Wang Y, Liu . Surgical treatment of skull fibrous dysplasia. *Surg Neurol* 2009;72: S17-20.
7. Boyce AM, Turner A, Watts L, Forestier-Zhang L, Underhill A, Pinedo-Villanueva R, Monsell F, Tessaris D, Burren C, Masi L, Hamdy N, Brandi ML, Chapurlat R, Collins MT, Kassim Javaid Muhammad. Improving patient outcomes in fibrous dysplasia/McCune-Albright syndrome: an international multidisciplinary workshop to inform an international partnership. *Arch Osteoporos* 2017;12:21.
8. Chapurlat RD. Medical therapy in adults with fibrous dysplasia of bone. *J Bone Miner Res* 2006;1 Suppl 2:P114-119.
9. Boyce AM, Kelly MH, Brillante BA, Kushner H, Wientroub S, Riminucci, M, Bianco M, Robey PG, Collins MT. A randomized, double blind, placebo-controlled trial of alendronate treatment for fibrous dysplasia of bone. *J Clin Endocrinol Metab* 2014;99:4133-4140.
10. Bousson V, Rey-Jouvin C, Laredo JD, Merrer ML,Martin-Duverneuil N, Feydy A, Aubert S, Chapurlat R, Orcel, P. Fibrous dysplasia and McCune-Albright syndrome: imaging for positive and differential diagnoses, prognosis, and follow-up guidelines. *Eur J Radiol*2014; 83:1828-42.
11. Efune G, Perez CL, Tong L, Rihani J, Batra PS. Paranasal sinus and skull base fibro-osseous lesions: when is biopsy indicated for diagnosis? *Int Forum Allergy Rhinol* 2012; 2:160-165.
12. Rafael Denadai, Cesar Augusto Raposo-Amaral, Frederico Figueiredo Marques, Enrico Ghizoni, Celso Luiz Buzzo, Cassio Eduardo Raposo-Amaral. Strategies for the Optimal Individualized Surgical Management of Craniofacial Fibrous Dysplasia. *Ann Plast Surg*2016;77:195-200.
13. Kusano T, Hirabayashi S, Eguchi T, Sugawara Y. Treatment strategies for fibrous dysplasia. *J Craniofac Surg* 2009; 20:768-770.
14. Ya-Ting Wei, Shu Jiang, Ying Cen. Fibrous dysplasia of skull. *J Craniofac Surg* 2010; 21:538-542.
15. Priya Jeyaraj. Histological diversity, diagnostic challenges, and surgical treatment strategies of fibrous dysplasia of upper and Mid-Thirds of the craniomaxillofacial complex. *Ann Maxillofac Surg* 2019;9:289-314.
16. Denadai R, Raposo-Amaral CA, Marques FF, Ghizoni, E, Buzzo CL, Raposo-Amaral CE. Strategies for the optimal individualized surgical management of craniofacial fibrous dysplasia. *Ann Plast Surg* 2016; 77:195-200.
17. Chan EK. Ethmoid fibrous dysplasia with anterior skull base and Intraorbital extension. *Ear Nose Throat J* 2005; 84:627-628.
18. Cutler CM, Lee JS, Butman JA, FitzGibbon EJ, Kelly MH, Brillante BA, Feuillan P, Robey PG, DuFresne CR, Collins MT. Long-term outcome of optic nerve encasement and optic nerve decompression in patients with fibrous dysplasia: risk factors for blindness and safety of observation. *Neurosurgery* 2006;59:1011-1017.
19. Satoh K, Mitsukawa N, Abe T. Is prophylactic decompression of the optic canal necessary in surgical correction of fronto-orbital fibrous dysplasia?: a review of 11 consecutive cases.*J Craniomaxillofac Surg* 2014; 42:1614-1616.
20. Amit M, Collins MT, FitzGibbon EJ, Butman JA, Fliss DM, Gil Z. Surgery versus watchful waiting in patients with craniofacial fibrous dysplasia-a meta-analysis. *PloS one* 2011; 6:e25179.
21. Holl DC, Hardillo JAU, Dammers R,van der Schroeff MP, van der Lugt A. Cystic Degeneration of Craniofacial Fibrous Dysplasia. *World Neurosurg* 2018; 120:159-162.
22. Liu Y, Yu H, Zhen H. Navigation-assisted, endonasal, endoscopic optic nerve decompression for the treatment of nontraumatic optic neuropathy. *J Craniomaxillofac Surg* 2019; 47:328-333.

Table 1. Clinical data of Observation Group Patients.

No.	**Type of disease**	**Age**	**Sex**	**Involved bones**	**With the paranasal sinu**
1	MFD	56	M	Sphenoid bone	
2	MFD	50	F	Sphenoid bone	Fungal maxillary sinusitis
3	MFD	34	F	Sphenoid bone	Chronic rhinosinusitis
4	MFD	42	F	Sphenoid bone	
5	MFD	11	M	Maxilla	

6	MFD	23	M	Perpendicular plate of ethmoid and middle turbinate	
7	MFD	49	F	Sphenoid bone	
8	MFD	67	M	Sphenoid bone	Chronic rhinosinusitis
9	MFD	26	F	Frontal bone	

MFD, monostotic fibrous dysplasia; FESS, functional endoscopic sinus surgery.

Table 2. Clinical Data of Patients Undergoing Surgery

No.	**Type of disease**	**Age**	**Sex**	**Involved bones**	**Associated disease**
1	MFD	55	M	Ethmoid bone	Chronic rhinosinusitis, headache
2	MFD	19	M	Ethmoid bone	Chronic rhinosinusitis
3	MFD	2	M	Ethmoid bone	Chronic maxillary sinusitis
4	MFD	33	F	Sphenoid bone	Necrotic polyps of right maxillary sinus, Cystic degeneration o
5	MFD	59	F	Sphenoid bone	Cystic degeneration, headache
6	MFD	18	M	Ethmoid bone	Chronic rhinosinusitis
7	MFD	22	F	Ethmoid bone	Headache
8	MFD	45	F	Sphenoid bone	Headache
9	MFD	54	M	Sphenoid bone	Headache
10	MFD	30	M	Sphenoid bone	Chronic rhinosinusitis

Table 3. Clinical Data of the Patients Receiving Endoscopic Optic Nerve Decompression (EOND)

No.	**Type of disease**	**Age**	**Sex**	**Involved bones**	**Treatment**	**Visual acuity Preop**	**Visual acuity Postop**
1	PFD	17	F	Maxillofacial; rib and humerus; sphenoid	Partial dissection and bilateral EOND	light perception in left eye, no light perception in right eye	0.7 0.3
2	PFD	55	M	Sphenoid, ethmoid	Partial dissection and right EOND	0.8	1.2
3	PFD	21	F	Sphenoid, ethmoid, and temporal; rib	Partial dissection and left EOND	0.5	1.0

4	McCune-Albright syndrome	10	F	Sphenoid, ethmoid, frontal, maxillary, zygoma, and temporal; rib, bilateral tibia and right fibula	Partial dissection and the left EOND	0.12	0.6

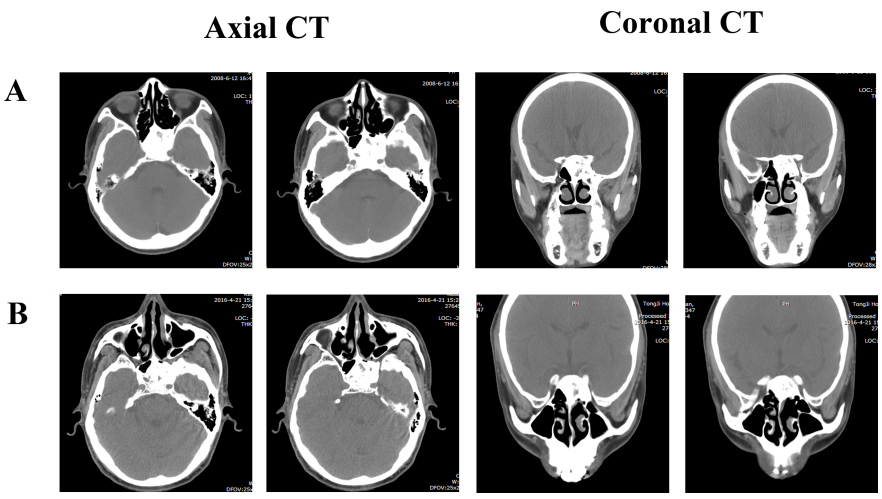
PFD, polyostotic fibrous dysplasia; EOND, Endoscopic Optic Nerve Decompression.

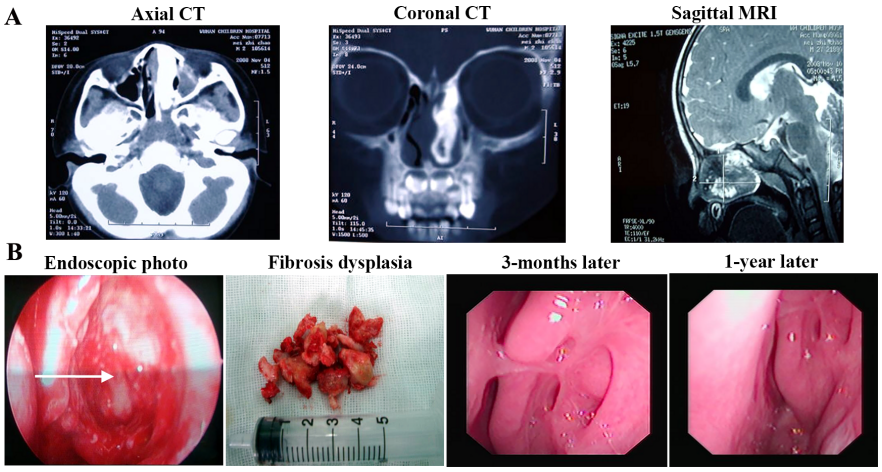
FIGURE LEGENDS

Fig 1. The imaging features of fibrous dysplasia located in the sphenoid bone in a 56-year-old patient (patient 1 in Table 1) in different dimensional CT scans (A). Because of no symptoms, the patient accepted a biopsy, which was identified as fibrous dysplasia. The patient was followed up until now. The image shows the nasal cavity and sinus of the patient eight years after the biopsy (B). (CT, computed tomography).

Fig 2. The imaging features of a 2-year-old patient (patient 3 in Table 2). The images show the axial and coronal views of CT scans and the sagittal view of the MRI of the paranasal sinuses before the operation (A). The removal of a fibrosis dysplasia lesion in the ethmoid bone under FESS (B). The mass removed from the ethmoid bone in the operation was confirmed to be fibrosis dysplasia by pathological examination (B). The photos under nasal endoscopy showed the nasal cavity after the endonasal endoscopic operation 3 months later and 1 year later, respectively (B). The white arrow shows the place of the removed lesion in the ethmoid bone. (MFD, monostotic fibrous dysplasia; CT, computed tomography; MRI, magnetic resonance imaging; FESS, functional endoscopic sinus surgery).

Fig 3. The imaging features of MAS in a ten-year-old patient (patient 4 in Table 3) (Fig. 3A and 3B). Images used during the navigation-assisted operation: sagittal, coronal, and axial scans, and the video (The first and the second operation: 3A and 3B). The patient received left optic nerve decompression using an endoscopic endonasal approach. Then the patient received left optic nerve decompression again one and a half years later because of her left visual loss again. The green dot shows the left orbital apex region (3A, 3B). Fig.3C showed the images used to aid image-guided navigation in MFD endoscopic sinus surgery (patient 9 in Table 2) . The white arrow shows the thin layer of fibrous tissue between the fibrous dysplasia lesions and the sphenoid bone (Fig.C). Fig.3D showed the imaging features of fibrous dysplasia with degeneration changes (patient 5 in Table 2). The paranasal sinus CT of MFD showed frosted glass changes of the left sphenoid pterygoid process and the great wing, with a 2.3× 1.0 cm soft tissue shadow, which was diagnosed as hyaline degeneration by pathological examination (Fig.3D). The CT scan showed the cystic degeneration of FD (Fig.3E-H). Axial CT scan of the 17-year-old patient (patient 1 in Table 3) with PFD showed the bilateral optic nerve compression due to cystic degeneration of FD (Fig.3E). The CT scan of two years before surgery revealed that the left optic nerve was completely encased and the right optic nerve was partially encased by FD (Fig.3F,G,H). (CT, computed tomography; MFD, monostotic fibrous dysplasia; PFD, polyostotic fibrous dysplasia; FD, fibrous dysplasia)





Photos in surgical navigation