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CASE REPORT

Fibrous dysplasia of occipital bone revealed by acute intracranial hypertension

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Abstract

Fibrous dysplasia of bone is a stem cell bone disease due to a somatic GNAS mutation which can affect craniofacial bones. Although craniofacial fibrous dysplasia is a benign and progressive disorder, it can cause mass effect on the cranial structures. We describe an 18-year-old man, without past medical history, came at the emergency department with progressively worsening headache, associated with vomiting. Cranial bone CT and then brain MRI revealed fibrous dysplasia of occipital bone with intraosseous cyst, compression of right sigmoid vein. An angiography was performed to stent the right sigmoid vein and symptoms had completely resolved only a few hours after the procedure. Then, a treatment by bisphosphonates was introduced. We believe this is the first description of sigmoid vein compression by a bone cyst, requiring stenting. MRI should be performed urgently in case of unusual severe headache or rapidly evolving neurologic impairment in patients with craniofacial fibrous dysplasia. Treatment of fibrous dysplasia is a controversial subject. In cases with neurologic complications, surgery or endovascular treatment might be performed.

Keywords Cranial bone · Fibrous dysplasia · Intracranial hypertension

Introduction

Fibrous dysplasia of the bone is a stem cell bone disease due to a somatic GNAS mutation which can affect craniofacial bones [2]. It is characterized by slow, progressive replacement of a localized area of the bone by an abnormal proliferation of isomorphic fibrous tissue intermixed with poorly formed, haphazardly arranged trabeculae of the woven bone [1]. Although craniofacial fibrous dysplasia is a nonmalignant and progressive disorder, it can cause mass effect on the cranial structures. Here, we describe an 18-year-old man with fibrous dysplasia of occipital bone revealed by intracranial hypertension.

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Observation

An 18-year-old man, without past medical history, came at the emergency department with progressively worsening headache, associated with vomiting. Brain CT with bone window and then brain MRI revealed fibrous dysplasia of the right occipital bone with intraosseous cyst, intra-cystic bleeding, and compression of the right sigmoid sinus. There was also a Chiari type 1 malformation (Fig. 1). Fibrous dysplasia also affected sphenoidal, C1, and C2 bones. Ophthalmological examination showed bilateral papillary edema grade I. Blood samples showed normal calcemia (2.31 mmol/L), phosphoremia (1.04 mmol/L), and renal function. Also, alkaline phosphatase was at 213 U/L ($N = 40–150$ U/L), CTX = 559 pg/mL ($N = 140–440$ pg/mL), and 25 OH vitamin D3 = 45 nmol/L ($N = 50–125$ nmol/mL). C-terminal FGF23 was normal, 90 Ru/mL ($N = 34–96$ Ru/mL). The patient was then hospitalized in the Neurology and treated by intravenous pamidronate 60 mg and mannitol 100 ml per 6 h.

The next day, the headache and vomiting aggravated, with impaired consciousness, bilateral VI palsy, and 4/5 motor deficit of the left hemi-body. A second brain MRI was performed but did not reveal additional abnormalities. Specifically, there

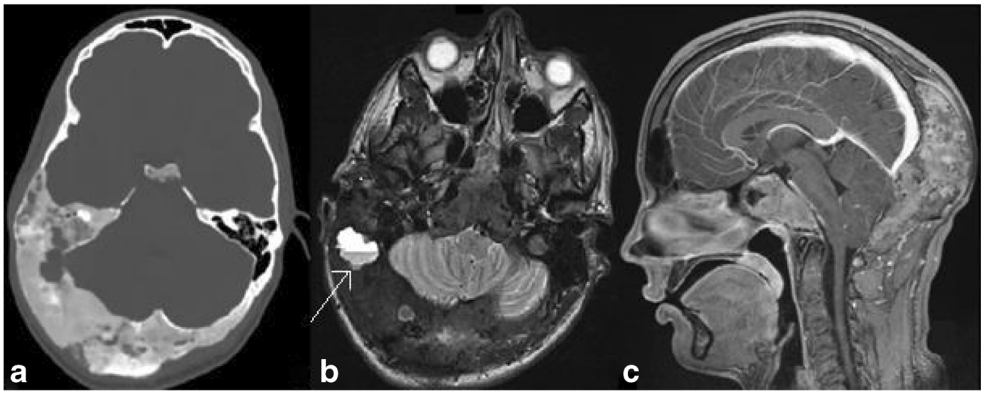

Fig. 1 **a** Cranial bone CT showing occipital bone fibrous dysplasia. **b**, **c** Brain MRI images of occipital bone fibrous dysplasia with intra-cystic bleeding (arrow), with spontaneous T1 hypersignal, T2 hyposignal, and with fluid level on brain MRI

was no hydrocephalus. Papillary edema increased on new ophthalmological examination. The patient was transferred to intensive care unit, and a cerebral angiography was performed and showed tight stenosis of the right sigmoid sinus, which could explain the signs and symptoms. (Fig. 2).

Two days later, after multidisciplinary discussion, a new cerebral angiography was performed to stent the right sigmoid sinus. Treatment with clopidogrel and acetylsalicylic acid was also introduced. Only a few hours after the procedure, symptoms had completely resolved. Visual monitoring did not find any abnormality. New brain IRM, performed 2 months later, exhibited a permeable stent in the sigmoid sinus, with good flow (Fig. 2).

In the following weeks, we explored the extension this previously unknown bone disease with bone scintigraphy, which showed uptake of cranial dysplasia area of the occipital, sphenoidal, and parieto-temporal right bones. There was no increased uptake of C1 and C2.

Six months after the diagnosis, the patient was asymptomatic and received a second course of pamidronate 60 mg per day for 3 days. Blood samples showed a normalization of markers of bone remodeling, with CTX = 324, bone alkaline phosphatase = 73, and FGF23 = 73.

Discussion

Craniofacial fibrous dysplasia of the bone is a well-described entity in which normal bone is replaced by abnormal fibroconnective tissue proliferation [1]. The clinical spectrum encompasses various forms including [2] the most common monostotic form (70%); the polyostotic form (30%); the McCune-Albright syndrome, a rare variant of with café-au-lait cutaneous spots and endocrine abnormalities; and the craniofacial forms that tend to be individualized because of potential neurologic and endocrine complications.

The frequency of the various craniofacial bones that can be affected by fibrous dysplasia of the bone remains controversial. Lustig et al. reported that mostly affected cranial bones were the ethmoid (71%), sphenoid (43%), frontal (33%), maxillary (29%), and the least common, the temporal (24%) and occipital bones (5%) [3]. In another study, it was reported that the frontal bones were most common involved followed by the sphenoid, ethmoid, parietal, temporal, and occipital bones [4]. Therefore, our patient had an uncommon presentation of a rare localization of cranial fibrous dysplasia.

We found eight other cases of occipital fibrous dysplasia in literature [5–12], systematically in young patients with a

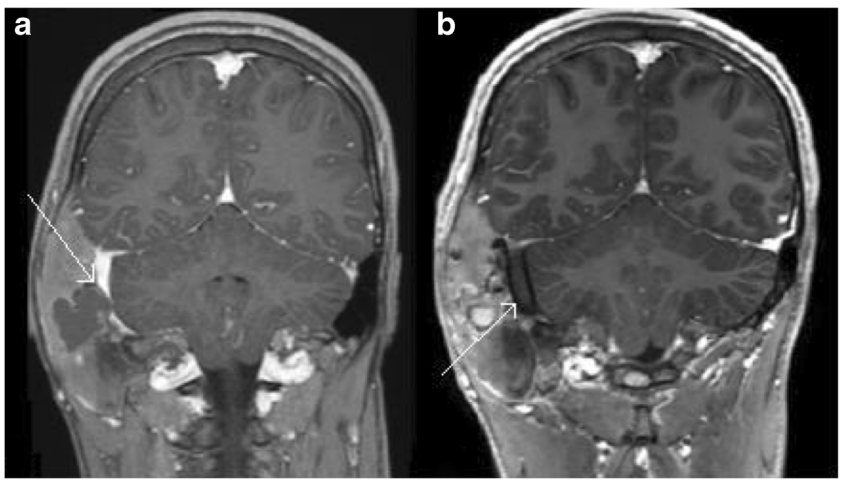

Fig. 2 **a** Brain MRI showing bone fibrous dysplasia with compression of right sigmoid sinus (arrow). **b** Brain MRI showing stenting of the right sigmoid sinus (arrow)

monostotic form of fibrous dysplasia. Most were revealed by an enlarging mass and only one by headache [8]. Our patient, although he noted an enlarging mass in recent years, presented uniquely a severe intracranial hypertension at the time of diagnosis.

For this patient, diagnosis was relatively easy with successive imaging techniques. CT scanning is the best way to display the bony changes. It also helps to distinguish fibrous dysplasia and various bone pathologies, e.g., Paget's disease. Brain MRI was especially used to evaluate the effect of fibrous dysplasia on adjacent tissue structures of the skull base [13].

The treatment of fibrous dysplasia is a controversial subject. In patients with bone pain, drugs that prevent osteoclastic bone resorption (bisphosphonates) can be used [14]. However, in the cases with neurologic complications, surgery might be performed [9, 11, 12]. In this case, our patient was treated by endovascular way and right sigmoid sinus stenting with fast and spectacular improvement. Then, treatment by bisphosphonates helped control bone pain. We believe the main lesson from this case is the potential for rapidly developing compression by growing cysts. In this case, the compression was of vascular nature, but it could also have been a direct neurologic compression affecting the optic nerve or another neurologic structure. MRI is of particular value for the diagnosis and should be considered in patients with craniofacial fibrous dysplasia who exhibit severe and rapidly changing symptoms.

Conclusion

We described for the first time a sigmoid sinus compression by a bone cyst, requiring stenting. MRI should be performed urgently in case of unusual severe headache or rapidly evolving neurologic impairment in patients with craniofacial fibrous dysplasia.

Compliance with ethical standards

Conflict of interest None.

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