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CASE REPORT Open Access

A case presentation of unilateral cavernous sinus inflammation due to monostotic fibrous dysplasia

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Abstract

Background: Craniofacial monostotic fibrous dysplasia is a rare and usually incidental bone pathology. CT comes to the fore in the diagnosis of monostotic fibrous dysplasia and MRI is used to show its relationship to the neighboring structures. Cavernous sinus inflammation may occur due to infectious and inflammatory causes. Inflammation of unknown cause is called Tolosa–Hunt syndrome.

Case presentation: In this case report, we presented cavernous sinus inflammation accompanying fibrous dysplasia in a 35-year-old female patient, which was seen for the first time in the literature.

Conclusions: The contrast-enhanced MR images are very important for the diagnosis and follow-up of the patient, in the rare presence of association we have shown in this case.

Keywords: Monostatic fibrous dysplasia, Cavernous sinus, Inflammation

Background

Fibrous dysplasia (FD) is a common, non-hereditary, genetic osteodystrophic disease. Impairment in skeletal stem cell function causes disruptions in osteoblastic activity and resorption. As a result, the normal bone structure is replaced by large fibrous stroma and islets of immature bone. Three main subtypes have been defined: monostotic, polyostotic, and McCune–Albright syndrome. Monostotic is the most common type (approximately %70 of patients) and is more common in adolescent females [1–3]. It frequently involves the long bones, ribs and craniofacial region. It shows predilection to the face and frontal bones within the skull bones and rarely occurs in the skull base and convexity. It is usually asymptomatic, whereas in symptomatic cases, the complaints are usually due to mass effect [4, 5]. Computed tomography (CT), which is sensitive to bone structure, is the first choice in diagnosis. Magnetic resonance imaging (MRI) is used to show its relationship with surrounding tissues and compression findings [6, 7].

Cavernous sinus syndrome (CSS) is defined as the deterioration of the functions of the anatomical structures in the cavernous sinus after any pathology affecting it. The most common cause is neoplasm, while inflammation takes the second place. While the most common cause is neoplasm, inflammation, vascular pathologies (cavernous sinus thrombosis, carotid–cavernous fistulas, carotid–cavernous aneurysms) and trauma are among other causes [8]. Sarcoidosis, Wegener's granulomatosis and IG4-related disease are specific causes of inflammation. Tolosa–Hunt syndrome is an idiopathic inflammation diagnosed after exclusion of other causes [9, 10]. Radiological imaging, especially MRI, plays an important role in the diagnosis and etiology of CSS. In the cavernous sinus, there is an increase in thickness, which causes convexity in the outer wall. It appears hypointense on T1-weighted images and iso-hypointense on

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T2-weighted images and shows contrast enhancement [10–12].

In this case, we aimed to show the rare association of ipsilateral cavernous sinus inflammation accompanying monostotic fibrous dysplasia in the right frontotemporal region.

Case report

A 35-year-old female patient applied to our clinic with a complaint of headache and right retro-orbital pain. The patient had no known chronic disease, no alcohol and smoking history. She had left stapedotomy surgery 2 years ago. Neurological examination did not reveal any specific features and the patient did not have any ocular findings. The patient underwent cranial CT without contrast. In the images, an expansile lesion was observed in the right frontotemporal bone, which caused an enlargement of the diploe distance consisting of widespread ground glass densities (Fig. 1). Monostotic fibrous dysplasia was initially considered as a preliminary diagnosis for the lesion causing destruction in the bony cortex. Contrast-enhanced brain MRI was performed to show the relationship of the lesion with cranial structures. In the right frontotemporal bone, intramedullary localized, lesion of approximately 54×23 mm, heterogeneously hypo-intense on T1-weighted images, heterogeneous hyper-intense on T2-weighted images was detected. On post-contrast images, heterogeneous enhancements in this area were accompanied by adjacent dural enhancement. MR images also showed enlargement of the right cavernous sinus. While this area is iso-hypo intense on T1-weighted images and isointense on T2-weighted images, the lesion had diffuse enhancement on post-contrast images (Fig. 2). Right frontotemporal craniectomy and cranioplasty were performed in the patient, whose possible other causes of cavernous sinus inflammation were excluded preoperatively. Histopathological diagnosis of FD was confirmed. The patient, who did not develop complications and whose complaints regressed, was discharged. When the patient came to follow-up approximately 1.5 years later, enlargement of the diploe distance and ground glass densities in the bone structure in the operation site were defined as recurrent-residual FD. Progression of cavernous sinus inflammation was detected in follow-up cranial MR images (Fig. 3). Reoperation was not planned and the patient was followed up with medical treatment.

Discussion

Head and neck bone involvement of monostotic FD accounts for approximately 50% of cases. Patients are usually asymptomatic and detected incidentally [13]. Although they show malignant transformation at a rate of less than 1%, follow-up is considered sufficient unless the patient has a significant complaint [14].

Cavernous sinuses are true dural venous sinuses and contain important structures. Compression-related symptoms occur in any pathology that may affect these structures, which are located at the base of the sphenoid bone and laterally with the temporal bone. Tolosa–Hunt

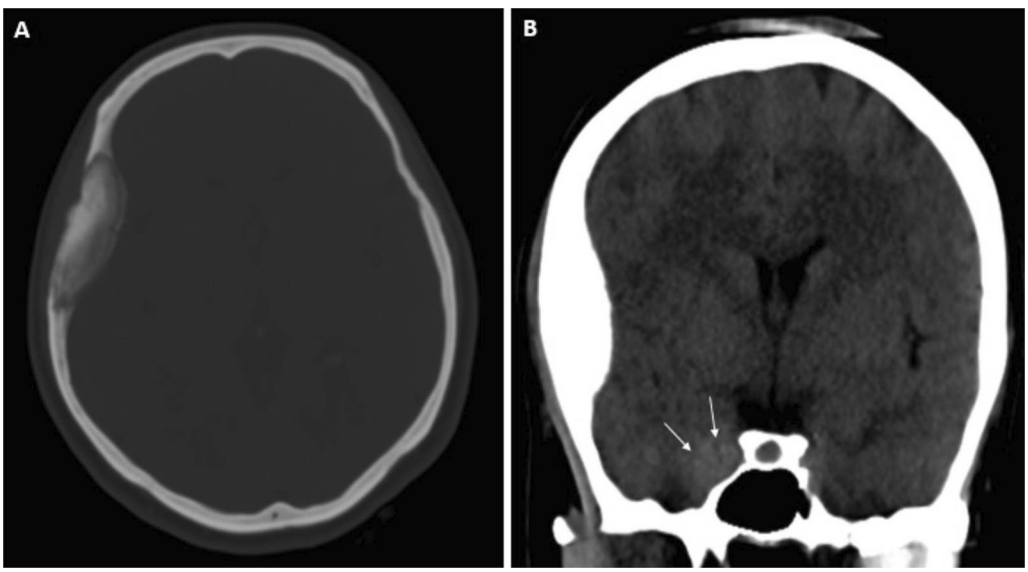

Fig. 1 In the axial section (**A**) of brain CT without contrast, in the right frontotemporal region an appearance of expansile, monostotic fibrous dysplasia is observed in ground glass density. In the soft tissue window of the coronal section (**B**), fullness in the right cavernous sinus draws attention (white arrows)

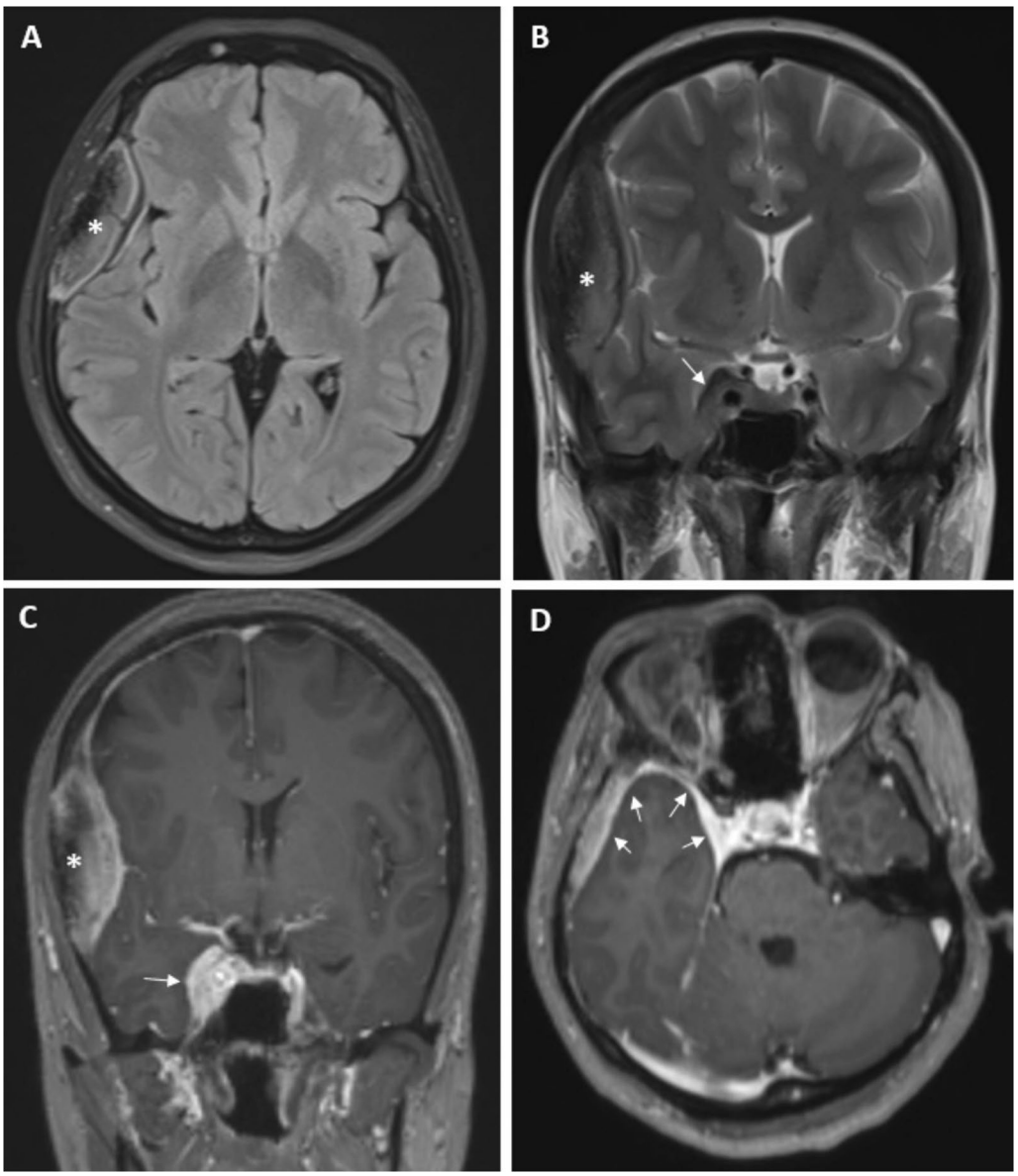

Fig. 2 In the FLAIR sequence, on the axial sectional brain MR image (**A**) and in the coronal section T2-weighted image (**B**), a right frontotemporal lesion of heterogeneous intensity located at the distance of the diploe is observed (asterisks). A hypo-isointense lesion filling the right cavernous sinus is seen in the coronal section (white arrow). Coronal (**C**) and axial (**D**) section post contrast T1-weighted images show the continuity of the enhancement in the bone lesion along the dura and homogeneous intense enhancement in the right cavernous sinus

syndrome is an idiopathic granulomatous inflammation of the cavernous sinus that may extend to the superior orbital fissure and orbital apex. The diagnosis of this painful ophthalmoplegic disease is made by clinical and neuro radiological imaging findings. Orbital or retro-orbital pain, nerve palsies, Horner's syndrome, symptoms that occasionally improve and remission, sometimes lasting for weeks, are among the common clinical features. It is known to respond dramatically to steroid therapy. The sensitivity of cranial CT in evaluating the cavernous sinus is low. Instead, MRI with high soft tissue resolution, especially coronal images, is a reliable diagnostic tool. The importance of MRI is to exclude other causes, especially mass lesion, that may cause CSS. Cavernous sinus filled with soft tissue is isointense with gray matter on T1-weighted images and iso-hypointense on T2-weighted images. The unilateral cavernous sinus is thickened and its lateral wall expands. Post-contrast

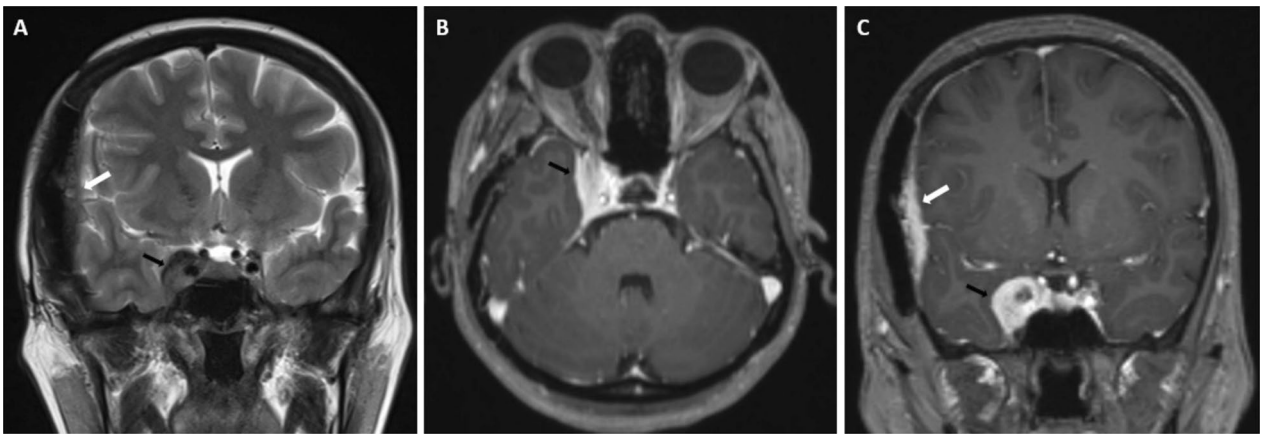

Fig. 3 Postoperative T2-weighted coronal (**A**), postcontrast axial (**B**) and coronal (**C**) images show a residual-relapse area of the lesion in the right frontotemporal region (white arrow). The expansion and enhancement of the right cavernous sinus appears to progress (black arrow)

images are diffusely enhanced. Narrowing of the carotid artery in the sinus can be seen [10, 15].

In our case, there was ipsilateral cavernous sinus inflammation accompanying FD, which was the first case we investigated in the literature. Both pathologies may have occurred sporadically. However, the continuation of dural enhancement adjacent to fibrous dysplasia with enhancement in the cavernous sinus on MRI suggested that these two conditions are more related. It can be predicted that FD causes inflammation in the dura, which progresses to the cavernous sinus. The progression of cavernous sinus inflammation in the case with post-operative residual FD may support this thesis.

Conclusions

Although monostotic FD is usually an incidental pathology, the accompanying cavernous sinus inflammation, as in our case, is valuable in terms of timely diagnosis and treatment of the patient. Cranial MR images with contrast are a reliable and effective method for the evaluation of adjacent dural enhancement and cavernous sinus structures.

Although very rare, on patients with monostotic FD, radiologist should carefully investigate cavernous sinuses for accompanying inflammation.

Abbreviations

FD: Fibrous dysplasia; CT: Computed tomography; MRI: Magnetic resonance imaging; CSS: Cavernous sinus syndrome.

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Author contributions

EG—conceptualization, data collection, writing initial draft. HO—writing initial draft, editing, supervision. All authors read and approved the final manuscript.

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Availability of data and materials

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Declarations

Ethics approval and consent to participate

Ethics committee approval was not considered necessary because it was a case report. Informed consent was obtained from the patient for this study.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Competing interests

The authors declare that they have no competing interests.

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