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Case Report

Calcifying fibrous tumor of the clivus presenting in an adult

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ABSTRACT

Calcifying fibrous tumor is a benign fibrous tumor. It rarely occurs in the clivus. The present study describes a case of a 56-year-old female, who was admitted to Taihe Hospital with dizziness not accompanied with headache for 2 months. Brain computed tomography examination revealed a well-defined, partially calcified lytic-expansile lesion in the clivus, which corresponded to an enhancing mass on contrast-enhanced magnetic resonance imaging. The patient underwent endoscopic resection. Subsequent pathologic examination of the resected tissue confirmed that the tumor was calcifying fibrous tumor. The patient was followed up for 3 months after operation without recurrence or metastasis.

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Introduction

Calcifying fibrous tumor (CFT) most commonly (not exclusively) involves the soft tissues, which was first described as a “childhood fibrous tumor with psammoma bodies” of peripheral axial soft tissue in 1988 by Rosenthal et al [1] and then, Fetsch et al [2] rename it as calcifying fibrous pseudotumor in 1993. CFT occurs most frequently in children and young

adults. The etiology and pathogenesis of CFT are controversial. Some people think it derived from inflammatory myofibroblastic tumors, however most studies have failed to support this. Ultrastructural studies revealed fibroblastic feature [3]. As previously reported [4], CFT mainly occurs in the stomach, small intestine, pleura, mesentery, and peritoneum. Another common location for CFT is the soft tissues of the extremities. CFT arising from the heart, mediastinum, lung, breast, or gallbladder is rare [5–9]. As far as we know, this is the first case

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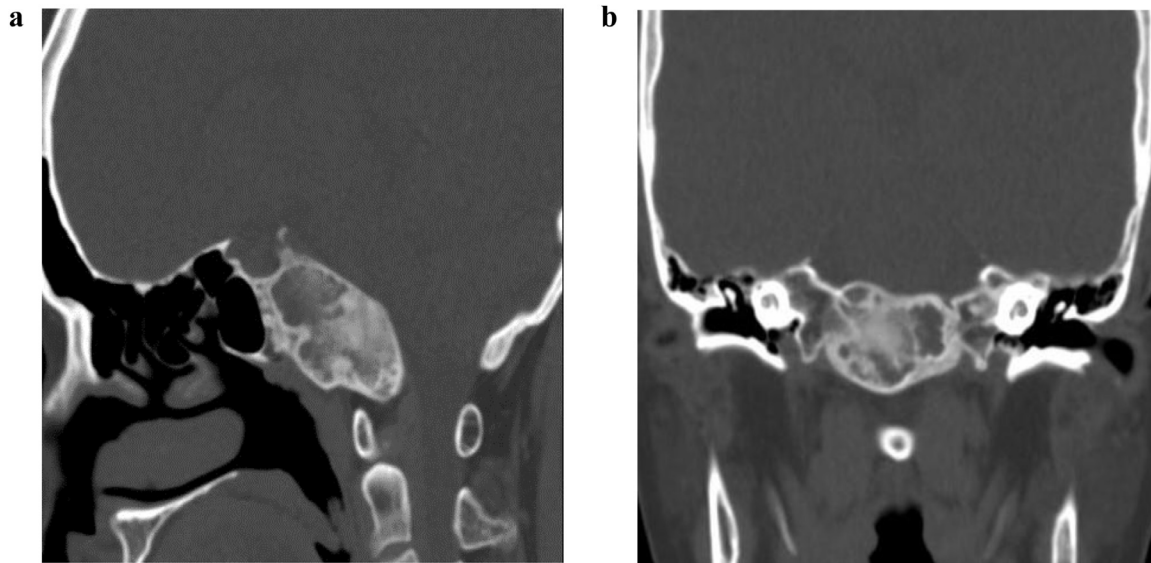


Fig. 1 – Sagittal (a) and coronal (b) CT images through the clivus (bone windows) demonstrate a well-defined, partially calcified, lytic-expansile lesion in the clivus.

of CFT that originated in clivus which was treated with endoscopic resection.

Case report

A 56-year-old female was admitted to our hospital with dizziness not accompanied with an obvious headache for 2 months, absence of nausea, vomiting, limb numbness, convulsion, or hoarseness. There was no relevant family or medical history. On physical examination no abnormality was detected. Laboratory work-up was unremarkable except for elevated blood cortisol level. Brain CT demonstrated a $40 \times 24 \times 31$ mm well-defined, partially calcified lytic-expansile lesion in the clivus, which corresponded to an enhancing mass on the delayed phase dynamic contrast-enhanced MR images. The lesion was diagnosed as neoplasia by CT and MRI which may be chordoma (Figs. 1 and 2).

As it was difficult to differentiate whether benign or malignant tumor, therefore endoscopic resection was carried out. Pathologic examination showed the tumor consisted of abundant hyalinized collagen tissue with lymphoplasmacytic mononuclear inflammatory cell infiltrate and dystrophic psammomatous calcifications (Fig. 3). Immunohistochemical examination revealed diffuse staining for Smooth Muscle Actin. All other antibodies cells tested were negative, including GFAP, EMA, PR, SSTR2, S-100, and CD34.

The patient recovered well after the operation, without recurrence or metastasis in a 3-month follow-up.

Discussion

CFT is rare entity that usually occurs in children and young adults. A slight increased tendency in women can be observed [4]. Its pathogenesis and mechanism have not been identified.

There is usually absence of characteristic symptom. Only diagnosed incidentally when there is certain increase in size. Elevated levels of IgG4 were often found in the serum, and positive plasma cells of IgG4 were found in CFT patients. Larson BK [10] hypothesized that CFT could be an IgG4-related disease based on observation of a high tissue content of IgG4-positive plasma cells.

Imaging may suggest the fibrous nature of the lesions. Sandy, spotty, and striped calcifications can be seen on CT and ultrasonography, as compared to x-ray examinations. The shape of the calcification may be variable during tumor growth. Ultrastructural studies show that calcification occurs as a result of cytoplasmic degeneration in the fibroblasts [11]. MRI can provide more valuable information about the lesion. On MRI, the tumor may be similar to fibromatoses [12], which shows hypointense signal on T1WI and T2WI. It may be due to being rich in hyalinized collagen fiber.

CFT should be distinguished from other tumors located in the clivus. The main consideration in the differential diagnosis of CFT in the clivus is chordoma. The classic radiological image shows a skull base chordoma which is a destructive soft tissue mass with speckles of bone within a minimally enhancing tumor on CT scan. On MRI the chordomas may be similar to CFT [13,14]. However, we can distinguish CFT from chordomas by histopathologically because cells with foamy and bubbly cytoplasm can be found in chordomas [15]. Since, their clinical manifestations and imaging findings are usually non-specific, histopathological examination is the main method to make a definitive diagnosis.

Microscopically CFT is characterized by dense hyalinization, fibrous proliferation, inflammatory infiltration by mononuclear cells, and scattered psammomatous and/or dystrophic calcifications [1]. The histopathological features of CFTs are generally easily distinguishable from other neoplastic lesions. The immunohistochemical examination does not play an important role in the diagnosis of CFT [16]. Fibroblasts

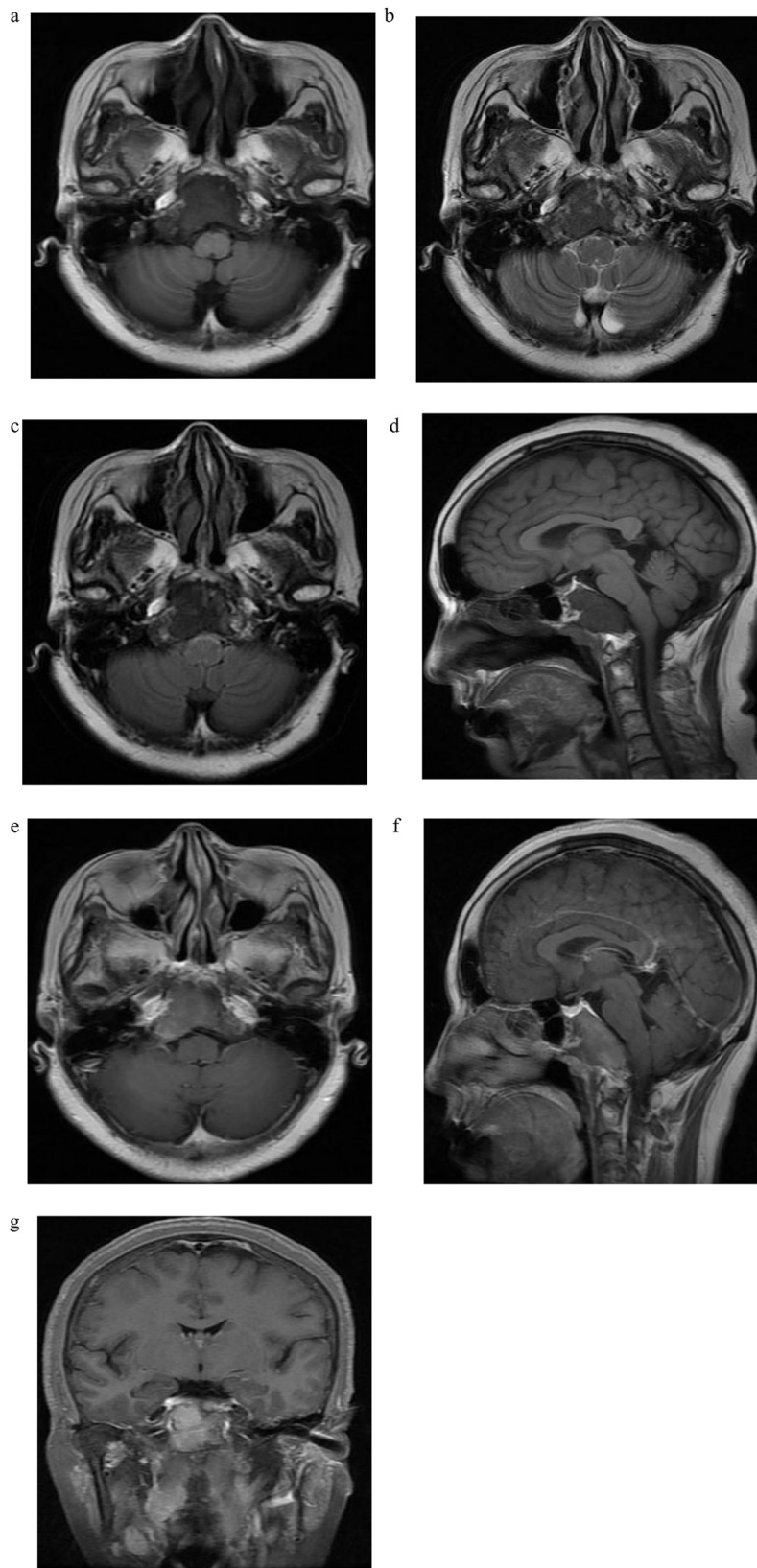


Fig. 2 - MR images through the clivus. The lesion demonstrates low signal intensity on axial (a) and sagittal (d) T1-weighted images and heterogeneous low signal intensity on the axial T2-weighted (b) and fluid attenuation inversion recovery (c) images. Axial (e), sagittal (f), and coronal (g) delayed-phase dynamic contrast-enhanced MR-images show the lesion enhances nonhomogeneously.

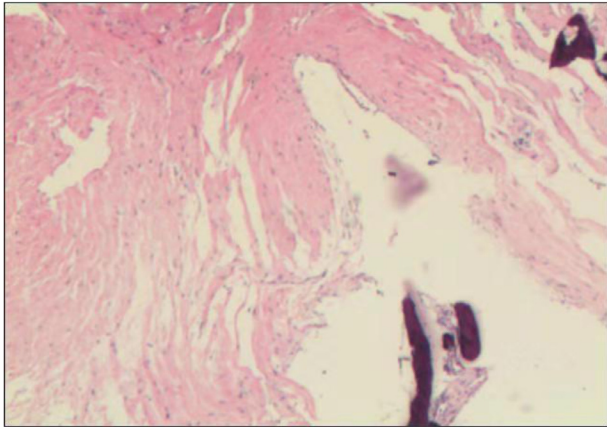


Fig. 3 – Hyalinized collagen tissue with lymphoplasmacytic mononuclear inflammatory cell infiltrate and dystrophic-psammomatous calcifications (H&E stain, x40).

do express vimentin. You can see the immune expression of factor XIIIa and CD34 on rare occasions [13,17].

CFTs have no tendency to metastasize. The treatment of CFT mainly includes the radical excision of the primary and possibly recurrent lesions, including open surgical excision and endoscopic resection. They are usually treated by local resection with clear margins. Nascimento et al [18] have observed local recurrences in 3 out of 10 patients in his study. Although the possibility of recurrence was little, re-excision should be taken into consideration [19]. Long-term observation and follow-up visits are necessary. Early diagnosis and effective treatment are the keys to the improvement of the clinical effects.

In summary, we conclude that CFT is a benign lesion that can occur in different parts of the body. In this case the characteristics of the lesions were studied postoperatively, based on histopathological examination. Imaging examinations have value in the diagnosis of CFT.

Supplementary material

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.03.028](https://doi.org/10.1016/j.radcr.2019.03.028).

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