

Case Report

Benign fibrous histiocytoma of the fronto-temporo-parietal region: a case report and review of the literature

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Abstract: Primary benign fibrous histiocytoma (BFH) at the skull is extremely rare. Here we report a case of a 22-year-old man presented with a 1-year history of progressive enlargement subcutaneous mass on the right side of the fronto-temporo-parietal region without symptoms. The tumor was radical resected through craniotomy and the bone defect was repaired by pre-plasticity titanium mesh. Histopathologic examination confirmed a benign fibrous histiocytoma, and no signs of tumor recurrence were detected at 3-year follow-up.

Keywords: Benign fibrous histiocytoma, skull neoplasm, neurocranium

Introduction

BFH, which consists of histiocytes and fibroblasts, is a benign mesenchymal origin neoplasm and most common affects the sun-exposed skin [1]. It accounts for approximately 1% of all benign bone tumors and extremely rare in the cranial bone [2, 3]. There are only eight cases have been reported in the literature [3-10]. Here we first report a case of an asymptomatic BFH involving the fronto-temporal and parietal bone regions in a man and review of the literature.

Case report

A 22-year-old man presented with a 1-year history of progressive enlargement subcutaneous mass on the right side of his head, which became soft gradually six months ago. It was interesting that the same place of his head got injury 2 years ago. However, clinical examination found neither pain nor neurologic deficits.

Neuroimage

Preoperative computed tomography (CT) images revealed an irregular osteolytic isodense mass lesion at the border of right frontal, tem-

poral and parietal bone regions, which eroded the entire thickness of the bone, and resulting in loss of both the internal and external bony plates of the skull. 3D reconstruction of skull showed the intricate architecture of the tumor (**Figure 1A-C**); On axial magnetic resonance imaging (MRI) scans disclosed a well-defined heterogeneous T1-isointense and T2-isointense lesion with focal areas of relatively hypointense and hyperintense respectively (**Figure 2A and 2B**). Axial and coronal Gd-enhanced T1-weighted images showed a homogeneously enhanced lesion with a dural tail sign. Intracranial extension of tumor had compressed the right frontal lobe without brain invasion (**Figure 2C and 2D**).

Operation

A right craniotomy was performed to excise the tumor under general anesthesia. A grayish-yellow rubbery lesion, which eroded the entire thickness of the skull and adhered to the subcutaneous tissue and outer layer of the dura mater slightly, was found during the operation. The tumor was well demarcated from the surrounding tissue and detached with little adhesion. Further resection was performed to remove damaged bone from the remaining normal structure of cortical bone, with completion

A case of skull neoplasm

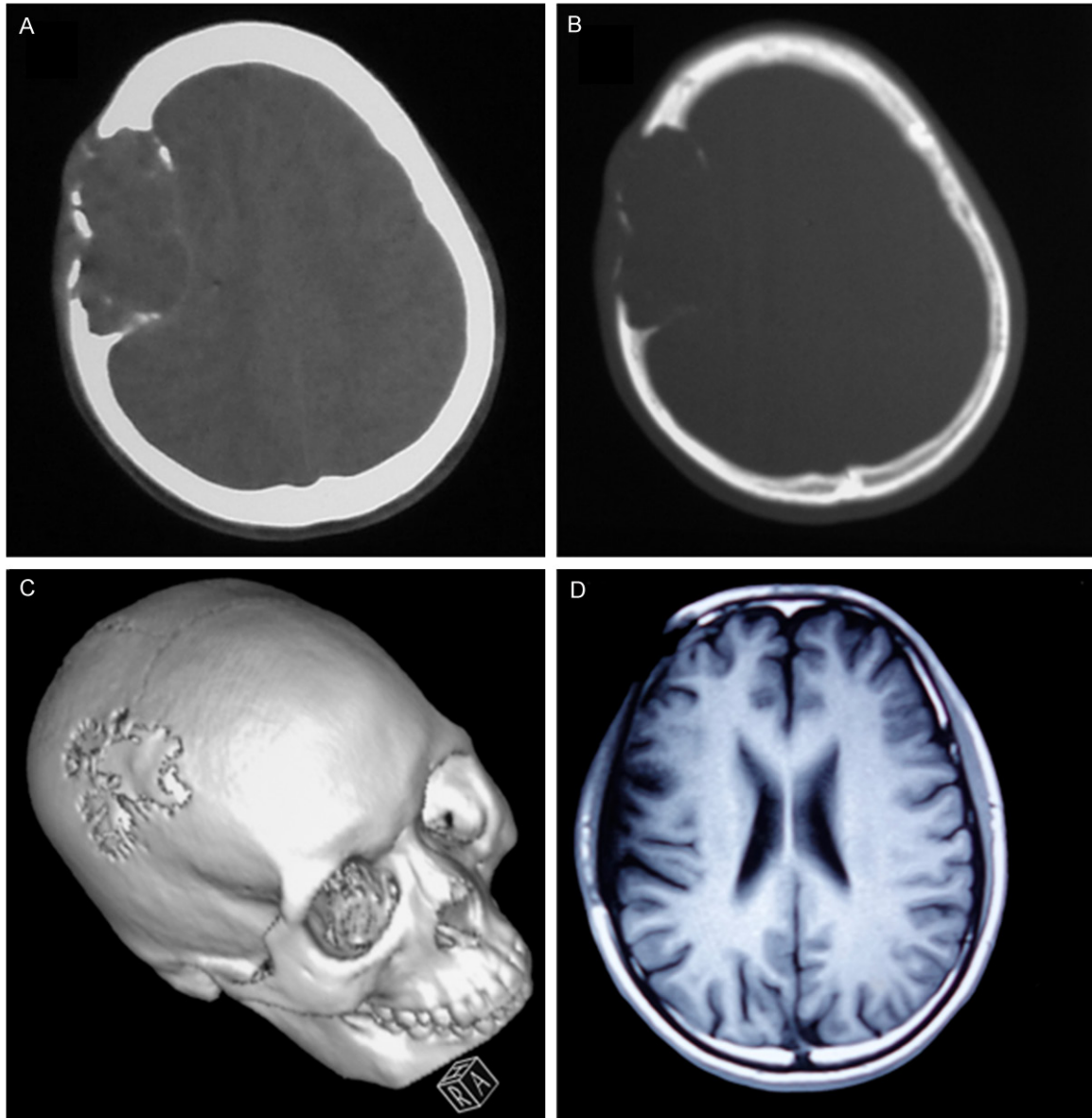


Figure 1. Preoperative brain CT and postoperative brain MRI. Preoperative brain-window CT image showed a iso-density mass with irregular shape (A), bone-window image disclosed the entire thickness of the bone eroded with a sclerotic border (B). A 3D reconstruction of skull demonstrated the intricate architecture of the tumor (C). Postoperative MRI scan of the axial showing no tumor recurrence 3-year after operation (D).

of repair using pre-plasticity titanium mesh. His post-operative course was uneventful.

Histopathology and follow-up

Postoperative histological examination demonstrated a proliferation of spindled-shaped tumor cells with some multinucleated giant cells and histiocytic foamy cells arranged in a storiform pattern (**Figure 3A**). There were no evidence of malignant characteristics include nuclear atypia, atypical mitosis, and necrosis.

Immunohistochemical analysis revealed that the tumor cells were positive for CD68 (**Figure 3B**) and negative for smooth muscle actin (SMA), desmin and CD34 (**Figure 3D-F**). There were some scattered S-100 positive cells (**Figure 3C**). The Ki-67 labeling index was few than 10% in most stainable areas. Histopathologic examination confirmed a benign fibrous histiocytoma, and no clinical or neuro-imaging signs of tumor recurrence and/or metastasis were found at 3-year follow-up.

A case of skull neoplasm

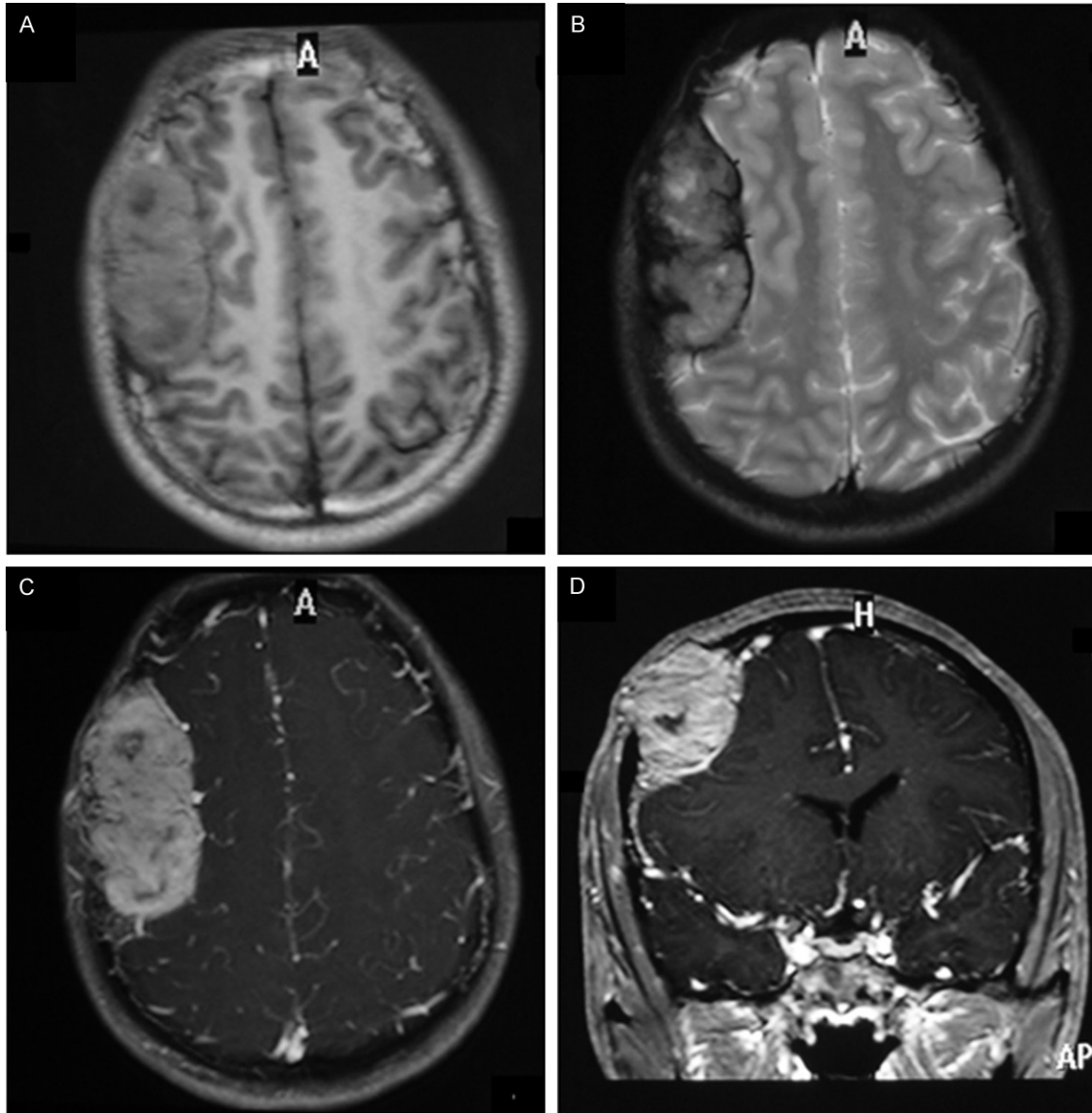


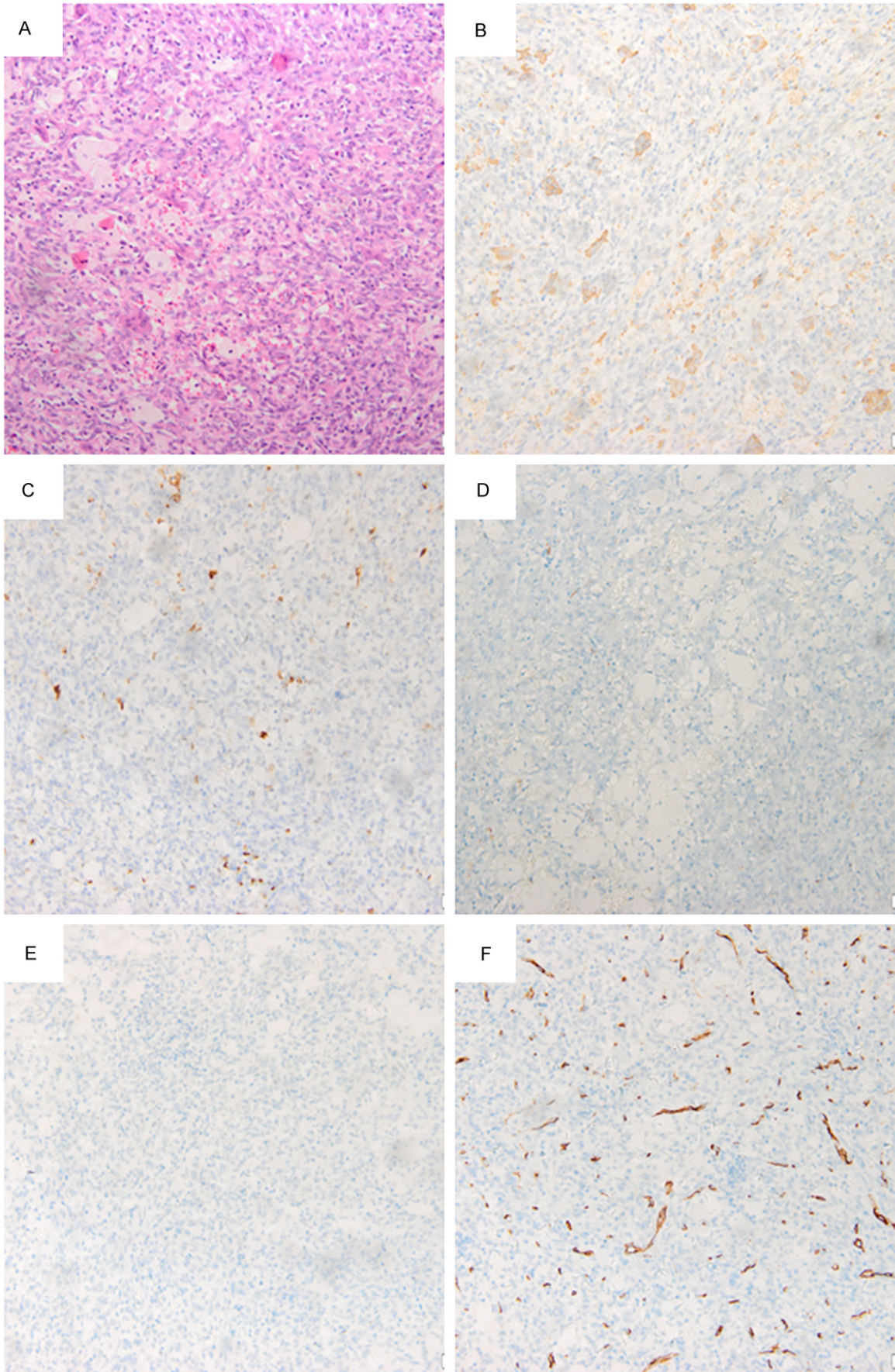
Figure 2. Preoperative brain MRI. On axial MRI scans disclosed a well-defined heterogeneous T1-isointense and T2-isointense lesion that compressed the right frontal lobe without brain invasion (A and B). Axial and coronal Gd-enhanced T1-weighted images showed a homogeneously enhanced lesion with dural tail sign (C and D).

Discussion

BFH is a benign mesenchymal origin neoplasm which consists of histiocytes and fibroblasts, and first described by Stout and Lattes in 1967 [1, 11]. This tumor commonly occurs in the sun-exposed skin but rarely involves the deep soft tissues of the head and neck, especially in the neurocranium [1, 12]. There are only eight cases of BFH on the neurocranium have been reported (Table 1). These lesions are more common in females (female-to-male ratio 2:1)

and prone to affect young patients at a mean age of 24 years (ranged from 11 months to 54 years) in the review of the literature. Two of the patients are infant. Two lesions located in the skull base, three in the temporal bone, one in frontal bone, one in parietal bone, one in occipital bone and our case in fronto-temporo-parietal bone (Convex-to-skull base ratio 7:2). The symptoms of these patients depend on the tumor location, four patients exhibited pain and the rest without symptoms.

A case of skull neoplasm



A case of skull neoplasm

Figure 3. Photomicrographs of BFH. H&E stained section of tumor showing a proliferation of spindled-shaped tumor cells with some multinucleated giant cells and histiocytic foamy cells arranged in a storiform pattern (A). Microphotographs of specimen histology showing the tumor cells positive for CD68 (B), partially positive for S-100 (C), negative for SMA (D), DESMIN (E), CD34 (F). Immunohistochemical staining, original magnification × 400 (A), × 200 (B-F).

Table 1. Clinical characteristics of benign fibrous histiocytoma at the skull

Author (year)	Age (years)/gender	Location	Symptoms	Operation	Follow-up	Recurrence
Fritz (2006)	45/M	Skull base	Pain/numbness	Near-total removal ¹	3 months	Yes (died)
Tubbs (2007)	11 months/F	Skull base	No	Biopsy	18 months	No
Ideguchi (2009)	33/F	Temporal bone	Pain and tinnitus	Total removal	6 months	No
J Wen (2010)	22/M	Frontal bone	exophthalmos	Near-total removal	6 months	No
Tsimiklis (2013)	25/F	Occipital bone	Pain	Total removal	1 years	No
Granato (2014)	56/F	Temporal bone	Pain	Total removal	2 years	No
Yang (2015)	12/F	Parietal bone	No	Total removal	12 months	No
Yamasaki (2015)	1/F	Temporal bone	No	Total removal	6 months	No
Present case	22/M	Frontal, temporal and parietal bone	No	Total removal	3 years	No

Preoperative imagings do great helpful to the diagnosis of BFH. On brain-window CT scan, BFH always shows a low density or isodensity radiolucent lesion with somewhat irregular edge and bone defects [10, 13]. BFH is typically osteolytic and localized with a sclerotic border on bone-window CT scans and homogeneous enhancement on CT with contrast [3]. In our case, CT scan showed thinning of the cortex as well as bone defects. There is a debate of the manifestation of this tumor on fluorodeoxyglucose (FDG)-PET. Some reported no specific uptake in patient with BFH, the others observed moderate or high FDG uptake on the contrary [10, 13, 14]. Most of the previously reported BFH of the skull were isointense on T1-weighted, relatively high-intense on T2-weighted and homogeneously gadolinium-enhanced. Ya-masaki et al. [3] detailed described BFH was low-perfusion on perfusion-weighted imaging (PWI) and iso-to-low intensity on diffusion-weighted imaging (DWI) scans, and it was helpful to distinguish BFH from malignant tumors because the later were high-intense on DWI due to restricted diffusion [15].

The differential diagnostic of BFH from acute inflammation, benign tumors include giant cell tumor, meningioma and ossifying fibroma and malignant lesions include malignant fibrous histiocytoma, fibrosarcoma, and osteosarcoma is very important. Radiologic manifestation can exclude the possibility of inflammation and Langerhans cell histiocytosis [3, 8]. The histopathological characteristics of BFH in bone include the proliferation of spindled-shaped

fibrohistiocytic tumor cells with some multinucleated giant cells and foam cells arranged in a storiform or swirl pattern [9]. Nuclear atypia, pleomorphism, atypical mitosis and necrosis that represent malignant characteristics are rarely found. BFH is usually positive for CD68, vimentin and α -SMA, and negative for CD34, CD1a, S-100, desmin and epithelial membrane antigens [16, 17]. These characteristics are conducive to distinguish BFH from other lesions. In our case, tumor cells arranged in a storiform pattern without malignant characteristics, and positive for CD68, negative for CD34, SMA and desmin with a Ki-67 labeling index few than 10%.

Surgery combined with radiation or chemotherapy can provide a good curative effect for many tumors. However, the best way to treat BFH in skull is surgical radical resection. The efficacy of adjuvant radio-chemotherapy after surgery is unknown. Up to two thirds of the patients may get local control in the postoperative radiotherapy [18]. Fritz et al. [4] reported a patient died of recurrence after radiotherapy following surgery. Chemotherapy in the majority of cases is invalid, but some cases achieve partial response after adriamycin or vinblastine [19, 20]. In the review of literature, an 11% recurrence rate after local excision in the head and neck was reported by Bielamowicz et al. [1]. Consequently, careful long-term clinically and radiologically follow-up is an important and effective way to monitor for tumor recurrence or metastasis.

Conclusion

In conclusion, this is the first case of primary BFH involves frontal, temporal and parietal bone regions at the same time. Radiological and histopathological manifestation can contribute to differentiate BFH from other lesions. Surgical total removal is the most effective way to treat it. Whether choose adjuvant radio-chemotherapy should depend on the specific condition of each patient. Long-term follow-up for monitoring tumor recurrence or metastasis is essential.

Disclosure of conflict of interest

None.

Authors' contributions

All authors were involved in clinical care and investigative workup of the patient. Hongxu Chen and Pengcheng Li provided pictures of the patient, drafted and revised the manuscript. Zhiyong Liu performed the patient follow-up, drafted and revised the manuscript. Jianguo Xu and Xuhui Hui were responsible for the study concept and revised the manuscript for intellectual content.

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