A solitary fibrous tumor in the cerebellar hemisphere parenchyma: a case report and literature review

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Abstract

Solitary fibrous tumors (SFTs) of the central nervous system occurring in the brain parenchyma are rare. We report an elderly female who presented with intermittent headaches and a solitary mass was noted in the right cerebellum that was misdiagnosed preoperatively as a hemangioblastoma. The postoperative pathologic diagnosis was a SFT. When an isolated mass appears in the cerebellar hemisphere with slight hyperdensity on CT images, heterogenous signals on T1WI and T2WI images, and a nodule with a hypointense signal on a T2WI image, the possibility of a SFT should be considered.

Keywords: isolated fibrous tumor, central nervous system, cerebellar hemisphere

1. INTRODUCTION

Solitary fibrous tumors (SFTs) of the central nervous system are rare, with an incidence of <1% [1, 2]. SFTs originate in the mesenchyma of the meninges. Central nervous system SFTs tend to occur in the meningeal region and are rare in the brain parenchyma, ventricles, and pineal gland [3, 4], with a subtentorial incidence of 34.4% [5]. The age of onset for SFTs is usually 40–70 years and the clinical manifestations are related to the anatomic site, such as headaches, numbness or convulsions of the limbs, and vision loss. SFTs are borderline tumors, most of which are benign, and 10%–20% are malignant or potentially malignant.

2. CASE REPORT

A 73-year-old female reported dizziness of 2 weeks duration without an apparent cause and intermittent seizures with nausea lasting 2–3 h that resolved spontaneously. She had no other complaints. Laboratory testing revealed no abnormalities.

A head CT pre-enhanced image showed a mass with mixed density in the right cerebellar hemisphere with slight hyperdensity, isodensity, and hypodensity inside (Figure 1a). MRI pre-enhanced images showed that the mass had mixed signals in T1WI and T2WI with a nodular presenting hypointense signal in T2WI and hyperintense signal in T1WI located at the posterior aspect of the lesion. The mass boundaries were clear. Mild edema surrounded the lesion and slight compression of adjacent structures was also noted. The lesion had a slightly hyperintense signal on DWI images (Figure 1b-d). MRI enhanced images showed heterogenous enhancement of the mass with a patchy area at the margin without enhancement representing cystic degeneration or necrosis. The nodularity at the posterior aspect of the lesion was mildly enhanced (Figure 1e). A 3D arterial spin labeling (ASL) image showed that hyperperfusion of the mass (Figure 1f). An MRS image showed that the choline peak (Cho) increased significantly and N-acetyl-aspartate (NAA) was absent (Figure 1g). The patient underwent radical tumor resection. The postoperative pathologic diagnosis was a central nervous system SFT (Figure 1h). The immunohistochemistry results were as follows: CD34 (+); CK (−); GFAP (−); S-100 (−); Ki-67 (5%+); PR (−); EMA (−); CD99 (weak +); NSE (−); Syn (−); vimentin (+); STAT6 (+); BCL-2 (+); CD31 (−); and...
Figure 1 | (a-i) Central nervous system SFT. (a) Head CT pre-enhanced image: a mixed density mass occurring in the right cerebellar hemisphere with slight hyperdensity, isodensity, and hypodensity. (b, c) MRI pre-enhanced images (b: T2WI, c: T1WI, d: DWI): the mass presented mixed signals on T1WI and T2WI images with a nodular presenting hypointense signal on T2WI images and hyperintense signals on T1WI images located at the posterior aspect of the lesion. The lesion had a slightly hyperintense signal on DWI images. (e) MRI enhanced image: the mass enhanced heterogeneously with a patchy area at the margin without enhancement representing cystic degeneration or necrosis. The nodularity at the posterior aspect of the lesion showed mild enhancement. (f) 3D ASL image: the mass had hyperperfusion. (g) MRS image: choline peak (Cho) increased significantly and N-acetyl-aspartate (NAA) was absent. (h) Pathologic picture: central nervous system SFT (HE staining, ×200). (i) MRI enhanced image approximately 16 months postoperatively: A new enhanced nodularity occurred at the area postoperatively.
Case Report

The tumor recurred at the surgical site 16 months postoperatively (Figure 1).

3. DISCUSSION

Central nervous system SFTs are rare, with an incidence of <1% [1, 2, 6]. Most reported intracranial cases originate from the dural or pia mater and rare arise from the brain parenchyma. In the current case the lesion occurred in the cerebellar hemisphere parenchyma. Between 10% and 20% of SFTs are malignant or potentially malignant and the prognosis is not strictly dependent on pathology. This case showed benign signs before surgery, but recurred 16 months after surgery, indicating that cases lacking malignant manifestations may also have aggressive potential, which is consistent with the previous literature [7].

The CT or MRI manifestations of the central nervous system SFTs are related to internal components, which can be summarized as follows in combination with the previous literature: (1) SFTs are circular or lobulated with clear boundaries. (2) Cystic degeneration, necrosis, or hemorrhage are common and calcifications are rare. (3) CT images usually show slight hyperdensity. MRI images show hypointense signals on T1WI, mixed signals on T2WI with curved, striped, patchy, or nodular hypointense signals ("Yin and Yang sign," "interweaving sign," or "black and white sign" [6]. (4) The hypointense signal area corresponds to numerous collagen fibers and sparse tumor cells, which are significantly enhanced, characteristic manifestations. The slightly hyperintense signal area corresponds to the area of tumor and vascular interstitial cells, and the enhancement is heterogeneous and moderate. The hyperintense signal area corresponds to the myxoid and necrotic areas and has no enhancement [6, 8]. Some scholars believe that lamellar or nodular hypointense signals on T2WI images are characteristic manifestations of SFTs [9]. In this case, a nodular hypointense signal on T2WI image existed. (5) DWI images show that the lesion has a hyperintense signal that is heterogeneous or ring-like [6]. (6) The tumor blood supply is abundant, and “empty blood vessel sign” in and around the tumor is common [6, 10]. No signs of “empty vascular flow” were noted in the current case. Some studies have defined >3 winding or tortuous blood vessels in tumors as a “serpentine sign” and these vessels do not originate from the same vessel. The “serpentine sign” is common in SFTs; the sensitivity and specificity are 81% and 92%, respectively [10]. (7) The “meningeal tail sign” is uncommon. (8) Peritumoral edema is associated with SFTs and the rate of venous sinus and skull invasion is high. In this case, mild peritumoral edema appeared around the lesion.

Central nervous system SFTs occurring in the infratentorial cerebellar hemisphere should be distinguished from hemangioblastomas, metastases, and ependymomas. Hemangioblastomas typically have a large capsule and small nodule, which is easy to diagnose. When the lesion is mixed with cystic and solid components, the lesion can be easily confused with other tumor lesions. The “empty vascular flow” inside and around the lesion and highly enhanced resembling blood vessel have value in differentiation. In the current case, there was no “empty vascular flow.” The degree of enhancement in the lesion is not as apparent as that of hemangioblastomas, so the two masses can be distinguished. Metastases in middle-aged and elderly people with a history of tumors, such as adenocarcinoma metastases, may show hypointense signals on T2WI images and are related to internal mucinous protein. Ependymomas occur in children and adolescents near the brain surface. Ependymomas are cystic and solid mixed, some of which have calcifications and bleeding with mild and strong enhancement. In the current case, the enhanced degree was more apparent than ependymomas. The slightly hyperdense area on CT images and hypointense signals on T2WI images with slight enhancement could exclude immature calcifications.

Central nervous system SFTs occur rarely in the sub-tentorial cerebellar hemisphere parenchyma. The possibility of SFTs should be considered when the isolated mass appears as hyperdense on CT images, mixed signals on T2WI images with nodular hypointense signals, and the characteristic “Yin and Yang sign.”

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DATA AVAILABILITY STATEMENT

The case is from The Affiliated Hospital of Hebei University.

ETHICS STATEMENT

This study was approved by the Ethics Committee of The Affiliated Hospital of Hebei University (Approval number: HDFY-LL-2019-042).

CONFLICT OF INTEREST

None.

REFERENCES


