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Case Report

Undetermined Fibrous Tumor with Calcification in the Cerebellopontine Angle

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In this report, we introduce an undetermined fibrous tumor with calcification occurring in the cerebellopontine angle (CPA). A 51-year-old woman was admitted with a short history of dizziness. Computed tomography and magnetic resonance images revealed a 2 × 2 × 2 cm sized mass at the left CPA which was round and calcified. There was no dura or internal auditory canal involvement. At surgery, the tumor was located at the exit of 7th and 8th cranial nerve complex. It was very firm, bright yellow and well encapsulated. Histologic findings revealed that the tumor was predominantly composed of fibrous component, scant spindle cells and dystrophic calcification. Immunohistochemical staining demonstrated positive for vimentin and negative for epithelial membrane antigen (EMA), S-100 protein, CD34, factor XIIIa and smooth muscle actin. The diagnosis was not compatible with meningioma, schwannoma, metastatic brain tumors, and other fibrous tumors. Although the tumor was resected in total, long term follow-up monitoring is necessary due to the possibility of recurrence.

KEY WORDS : Calcification · Cerebellopontine angle · Immunohistochemistry · Tumor.

INTRODUCTION

Intracranial tumors with calcification, which are present at cerebellopontine angle (CPA), consist of various benign and malignant tumors including meningioma, schwannoma, malignant glioma, metastasis and solitary fibrous tumors (SFT)[1,2,4,10]. Preoperative diagnosis is done by computed tomography, magnetic resonance (MR) images and thallium-201 SPECT which show dural involvement, bony erosion, proliferation potential and infiltration pattern to the normal parenchyma[8,23]. Differential diagnosis is a critical issue because the tumor can be treated not only by surgical excision but also with radiosurgery, conventional radiotherapy depending on clinical and radiological features[16]. However, it is sometimes difficult to determine the diagnosis and thus should be confirmed by the histopathologic examination. Here, we present a rare case of fibrous tumor with calcification which was located at left CPA. Although the tumor was resected in total, long-term follow up monitoring is necessary for the possible recurrence.

CASE REPORT

A 51-year-old woman was admitted with a history of dizziness for several months. She did not show any hearing impairment, facial palsy or cerebellar signs. Computed tomography (CT) revealed a 2 × 2 × 2 cm sized mass in the left CPA. Thallium-201 SPECT did not show thallium uptake increase in tumor compared to contralateral cerebellum (data not shown). There was no electrophysiologic evidence of facial neuropathy and audiogram resulted in normal range. In MR images, the tumor was hypointense signal on T2-weighted image and isointense on T1-weighted image with minimal contrast enhancement (Fig. 1). In addition, there was no contrast enhancement of the dura including left tentorium cerebelli. Furthermore, it seemed not to be related to the lower cranial nerves. At surgery, we identified that the tumor was very firm, bright yellow and well encapsulated round mass. It was also not adherent to the adjacent dura mater. The tumor was completely resected via a left suboccipital approach. After removal, there was small arachnoid adhesion at root exit region of 7th and 8th cranial nerve complex but no connection

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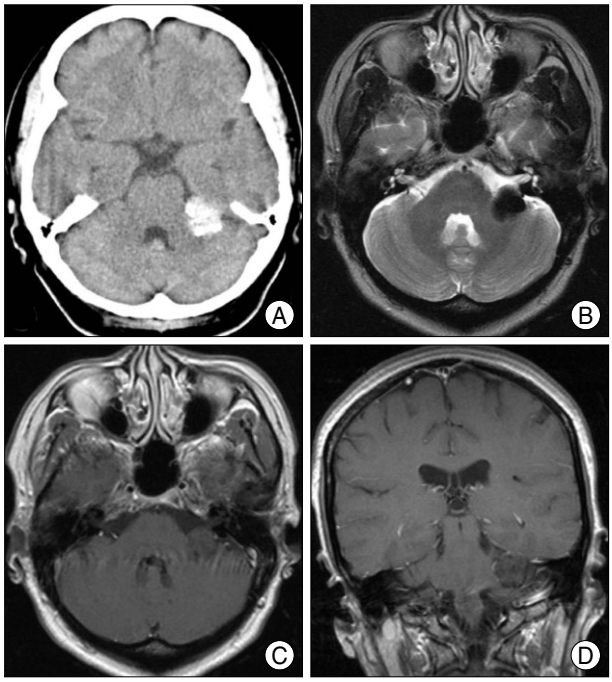

Fig. 1. In computed tomography, calcifying mass is seen at left cerebellopontine angle region (A) and T2-weighted magnetic resonance (MR) image shows hypointense lesion on left cerebellopontine angle (CPA) (B). The axial (C) and coronal (D) gadolinium enhancement MR images show isointense lesion with minimal enhancement on left CPA and there is no dural enhancement or internal auditory involvement of the tumor.

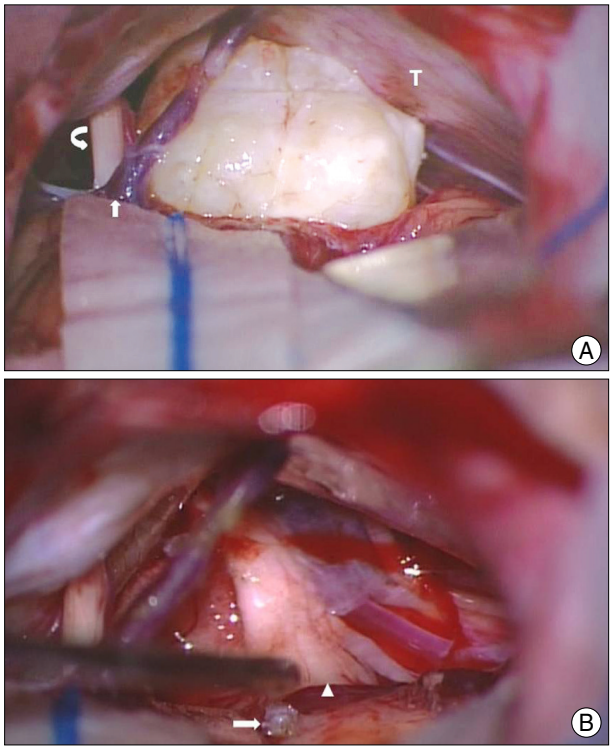

Fig. 2. The tumor is bright yellow and well circumscribed and it is not adhered to tentorium (T). A : Supeior petrosal vein (arrow) and 9th nerve (curved arrow) are observed. B : After tumor removal, arachnoid adhesion (arrow) adjacent to the exit of the 7th and 8th nerve complex (arrow head) is observed.

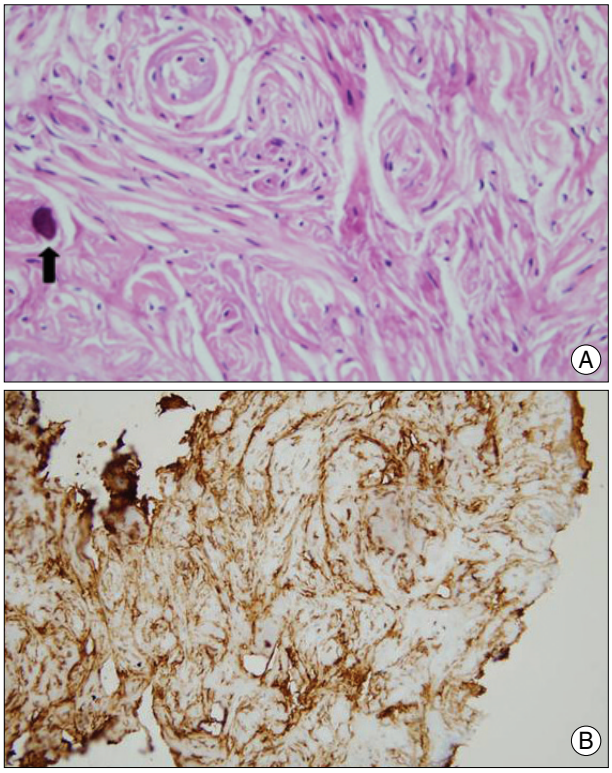

Fig. 3. Histopathological examination shows dystrophic calcification (arrow) and spindle cells (H&E, × 400) (A). Immunohistochemistry for vimentin is positive (B).

with these cranial nerves (Fig. 2). Histopathologically, the tumor was predominantly composed of fibrous component, scant spindle cells and dystrophic calcification. Immunohistochemical staining demonstrated positive for vimentin and negative for epithelial membrane antigen (EMA), S-100 protein, CD34, factor XIIIa and smooth muscle actin (Fig. 3).

The postoperative course was uneventful and 6 months follow-up MR images did not show remnant tumor or recurrence (Fig. 4).

DISCUSSION

Considering CT and MR images that the tumor was located in extraaxial CPA region, main differential diagnosis included meningioma, schwannoma and rarely metastatic tumors at first.

Meningioma is usually originated from arachnoid meningothelial cells and the dural membrane involving tumor shows strong contrast enhancement in MR images, although isolated meningioma can rarely be seen[24]. Histopathologically, meningiomas are characterized by whorls of cells, nuclear pseudoinclusions and psammoma bodies. It can be immunostained for EMA, and sometimes S-100. In addition, meningioma may express vimentin but only limited and locally positive for CD 34[3]. However, in our case, there was no dural enhan-

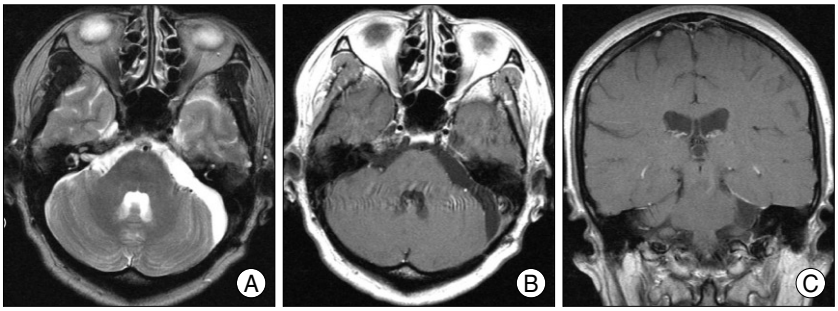

Fig. 4. T2-weighted (A) and gadolium enhanced T1-weighted (B and C) MR images checked 6 months after the surgery show no remnant or recurrence.

cement in MR images and arachnoid adhesion was only found at the exit of 7th and 8th cranial nerve complex interoperatively. In pathological examination, there were no meningioma cells, and the tumor was only positive for vimentin and negative for EMA, S-100, CD34 as well. Therefore, we could not confirm it as a meningioma. In case of schwannoma, the cisternal portion is typically larger than the intracanlicular portion suggesting the appearance of ice cream on a cone in MR images. In addition, the tumor composes of spindle-shaped neoplastic schwann cells including compact, elongated cells with occasional nuclear palisading (Antoni A pattern) and less cellular loosely textured or lipidized areas (Antoni B pattern). Furthermore, immunohistochemical staining demonstrates that S-100 is diffusely positive in schwannoma and CD34 positivity may occur in 89%. However, there was no intracanalicular involvement of the tumor in our case and the pathological features were not compatible with schwannoma[3]. There was no increased thallium uptake or malignant tumor cells in this case.

As the characteristics of fibrous tumor, solitary fibrous tumor (SFT) can be considered. SFT have been reported subsequently in virtually any body regions but rarely within the central nervous system since it was initially described as primary neoplasms of the pleura[5,13,25]. On MR imaging, the SFT is isointense with normal brain parenchyma on T1-weighted images, hyperintense on T2-weighted MR images, and shows homogeneous enhancement after intravenous administration of gadolinium[26]. In addition, patchy or focal areas of hypointensity and hyperintensity on T1- and T2-weighted imaging are notable findings in this tumor[12]. However, the SFT can sometimes be isointense to brain parenchyma on both T1- and T2-weighted MR images[14]. On histopathological features, SFT can be similar to fibrous meningioma or hemangiopericytoma (HPC). All of three tumors show spindle cells associated with collagen deposition and "stag-horn" vessel can be seen in both SFT and HPC[7,22]. In immunohistochemistry, SFTs show strong positivity for CD34, bcl-2, vimentin and negativity for EMA and S-100[22]. These findings may be similar pattern in our case which was negative for EMA, S-100 and positive for vimentin. However, CD34 staining was negative in the presenting case. Therefore, we could not confirm it as SFT.

There is one similar pattern to calcifying fibrous pseudotumor (CFP) in our case. CFP is a rare, benign tumor with a predilection for children and young adults that typically presents as a circumscribed nodule in subcutaneous or deep soft tissues[6] or in other visceral sites. It is histologically characterized by a cicatricose lesion composed of thick hyalinized collagenous, fibrous tissue including scanty spindle-shaped cells with psammomatous or dystrophic calcifications[15,21]. In immunohistochemistry, CFP is ordinarily positive for vimentin, factor XIIIa and CD68 and negative for smooth muscle actin, musclespecific actin, and CD 34[9,11,18,19]. However, in our case, there was no lymphoplasmocytic inflammatory cell infiltration and the tumor was negative for factor XIIIa staining. In addition, we could not found any report for CFT which was originated from CPA.

CONCLUSION

We report a rare case of surgically removed CPA fibrous tumor with calcification which was not determined in histopathologic examination. Although the tumor demonstrated benign characteristics in this case, long term follow-up should be done because most of tumor described can recur[3,7,17,20].

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