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Case Report

Intra-Suprasellar Schwannoma Originating from the Diaphragma Sellae

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A 49-year-old woman presented with headache, vomiting and visual disturbance. Neurological examination revealed bitemporal hemianopsia with poor visual acuity. Magnetic resonance imaging showed a bulky intra-suprasellar mass, which was isointense with brain parenchyma on T1-weighted images, and slightly hyperintense on T2-weighted images. After gadolinium administration, the mass was homogeneously enhanced. The mass was partially removed by the endonasal transsphenoidal approach and then the remnant mass was totally removed by the transcranial approach five months later. We found a yellowish mass which was attached to the diaphragm sellae in operation field. Histopathological examination of the tumor revealed the characteristic features of a schwannoma. We report an unusual case of an intra-suprasellar schwannoma resembling a non-functioning pituitary macroadenoma both clinically and radiologically.

KEY WORDS : Intrasellar · Schwannoma · Diaphragma sellae.

INTRODUCTION

Intracranial schwannomas account for 8-10% of all primary brain neoplasms[12]. Although sensory nerves are the preferred sites of development, motor and autonomic nerves may also be affected[17]. The vestibulocochlear nerve is the most frequently affected, and less commonly the fifth, ninth, tenth and seventh cranial nerves; as a result, 80 to 90% of cerebellopontine angle tumors are schwannomas[17]. However, schwannomas localized to intra-suprasellar area is extremely rare. We report an unusual case of an intra-suprasellar schwannoma resembling a non-functioning pituitary macroadenoma both clinically and radiologically.

CASE REPORT

A 49-year-old woman presented with headache, vomiting and visual disturbance. Neurological examination revealed bitemporal hemianopsia with poor visual acuity. The routine endocrine testing showed normal pituitary hormonal function. Computed tomography showed a snowman shaped mass lesion at the sellar region with upward displacement of optic chiasm and erosion of the sellar floor. Magnetic resonance imaging showed a bulky intra-suprasellar mass, which was isointense with brain parenchyma on T1-weighted images, and slightly hyperintense on T2-weighted images (Fig. 1A, B). After gadolinium administration, the mass was homogeneously enhanced (Fig. 1C, D). A provisional diagnosis of a non-functioning pituitary adenoma was made.

The patient underwent surgery by a endonasal transsphenoidal approach. The mass was yellowish, hemorrhagic, elastic-in-consistency and hard, and it was partially removed by means of curettage and suction. The lesion radiologically resembled a pituitary adenoma, but intraoperatively it was suspected as having a different histological nature. The histopathologic examination showed nuclear palisading and whorl of the cells, and Verocay bodies corresponding to schwannoma. Five months later, the remnant mass was totally removed by the transcranial approach. Intraoperatively, a yellowish soft mass was adherent to the diaphragm sellae without extension into the cavernous sinus; multiple nerves were found between the tumor mass and diaphragm sellae. Therefore, this mass could have originated from the nerves that innervated the diaphragm sellae. Histopathological examination of the tumor revealed the characteristic

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features of a schwannoma. The tissue was composed of Antoni A and Antoni B fibers (Fig. 2A). Immunohistochemically, there was high and diffuse positivity for the S-100 protein (Fig. 2B). Also, the mass attached to the diaphragm sellae showed diffuse positivity for S-100 protein. (Fig. 2C).

Last follow-up postoperative magnetic resonance imaging showed no remnant mass and recurrance. (Fig. 3).

DISCUSSION

Schwannomas account for about 8% of all primary intracranial neoplasms and are relatively frequent intracranial tumors. They usually develop from sensory nerves, most often the auditory nerve and less commonly the trigeminal nerve, although they have been shown to involve all other cranial nerves except the optic nerve[11,13,14,18]. However, an intrasellar location is exceptional and can mimick a nonfunctioning pituitary adenoma[1,3-5,8,9]. Schwannomas occurring within the sella are exceedingly rare; there have been 10 reported cases including the present case[6]. These tumors are not usually included in the differential diagnosis of sellar or suprasellar lesions. The clinical and radiological presentation of intrasellar schwannomas is consistent with the findings of pituitary adenomas. Therefore, many surgeons chose transsphenoidal approach as initial surgical procedure. Schwannomas have been described as fibrous, moderately to highly vascular and difficult to remove[3]. Hence, tumor removal is often incomplete because of the macroscopic features. Wilberger[16] recommended that a trans-sphenoidal approach should be avoided if a schwannoma is suspected. However, it is difficult to diagnose a sellar schwannoma preoperatively, because of their clinical and radiological resemblance to typical pituitary adenomas.

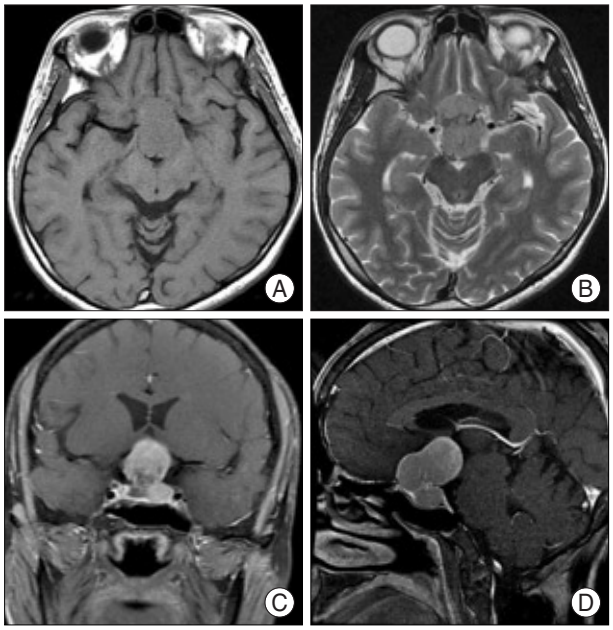

Fig. 1. Preoperative MR images. Magnetic resonance imaging shows a bulky intra-suprasellar mass, which is isointense with brain parenchyma on T1-weighted image (A) and slightly hyperintense on T2-weighted image (B). After gadolinium administration, the mass is homogeneously enhanced on coronal and sagittal images (C and D).

As there are no nerves observed within the sellar turcica, the origin of primary intrasellar schwannomas remains unclear. Three histopathogenetic hypotheses, which were about the origin from lateral sellar nerve plexus[1], perivascular schwann cells[4,9,12], and small sensory nerves that innervate the dura[3,5] have been proposed. Firstly, lateral sellar nerve plexus is a distribution centre for visceromotor and sensory nerves, which innervate cerebral arteries, orbital structures, and the dura mater[1]. The potential deficiency of the medial wall of the cavernous sinus has also been increasingly appreciated[8]. Secondly, Penfield[10] demonstrated the presence of perivascular schwann cells. Cerebral arteries as small as 10 to 15 μm in diameter receive adrenergic supply. The occurrence of schwannomas within the substance of the spinal cord has been attributed to the proliferation of schwann cells from nerve plexuses accompanying the perivascular spaces of perforating branches. In the third place, an origin from schwann cells that encircle small dural sensory branches of the trigeminal nerve or vasomotor nerves

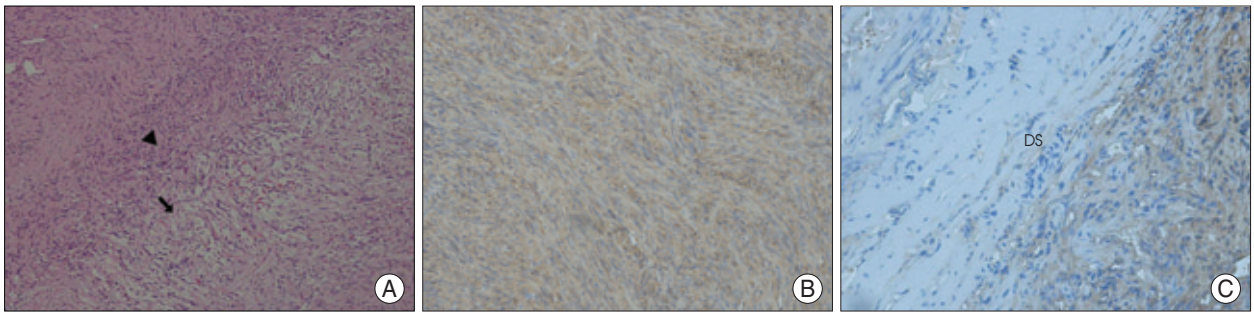

Fig. 2. Pathological findings. The tissue is composed of Antoni A and Antoni B fibers (A, H & E stain, original magnification × 100). Immunohistochemically, there is high and diffuse positivity for the S-100 protein (B, original magnification × 100). The mass attached to the diaphragm sellae also shows diffuse positivity for S-100 protein (C, original magnification × 200). Arrow : Antoni B fibers, Arrow head : Antoni A fibers, DS : diaphragm sellae.

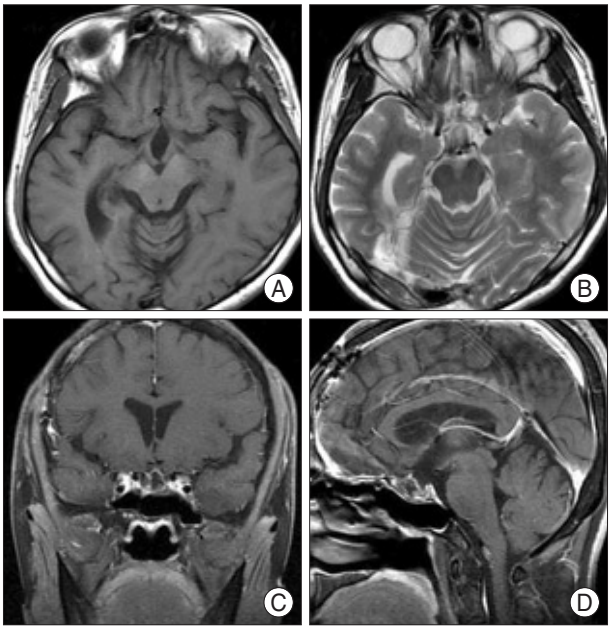

Fig. 3. Postoperative MR images. Last follow-up postoperative magnetic resonance imaging shows no remnant mass and recurrence.

has been proposed.

Intraoperatively, our case showed that the tumor had dense dural attachments adjacent to the diaphragm sellae. This mass might have originated from nerves that innervated the diaphragm sellae and the histopathological findings also suggested this origin of the tumor.

The pathological differential diagnosis for a sellar schwannoma includes three common lesions that can be observed in this region, namely fibroblastic meningioma, astrocytoma of the posterior pituitary gland and solitary fibrous tumors[7,12,18]. All four lesions including schwannoma have similar histological appearance as low-grade spindle cell tumors with cells arranged in fascicles. Therefore, immunohistochemical examinations are essential for confirmation of the diagnosis[18]. Fibroblastic meningiomas show moderate focal positivity for S-100, while schwannomas are diffusely and strongly reactive to S-100[7]. In addition, meningiomas are epithelial membrane antigen-positive, whereas schwannomas are negative. Astrocytomas of the posterior pituitary gland, which are also known as pituicytomas, are mostly immunopositive for glial fibrillary acidic protein (GFAP) and may demonstrate S-100 immunopositivity, similar to schwannomas[2]. A solitary fibrous tumor (SFT) has clearly different findings of the vascular stroma and the cellular appearance[15]. Furthermore, the immunohistological profile with positivity for CD34 is consistent with a SFT, whereas CD34 reactions are reported only in a very small portion of certain schwannomas. Our case showed immunopositivity to only S-100, not to GFAP, epithelial membrane antigen (EMA) and CD34. These findings confirm our case to be a schwannoma.

CONCLUSION

Authors experienced a very rare case of schwannoma in the sellar and suprasellar area originating from the small nerves that innervating diaphragm sellae. Because of the similarity with pituitary adenoma in the clinical and radiological aspect, it should be included in the differential diagnosis of sellar and suprasellar leaions.

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