

Primary Intrasellar Schwannoma - A Case Report -

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Received : February 15, 2002
Accepted : April 29, 2002

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Primary intrasellar schwannomas or neurilemmomas occur rarely and mimic pituitary adenoma, radiologically and clinically. The authors describe the 6th case of primary intrasellar schwannoma mimicking a nonfunctioning pituitary macroadenoma, clinically as well as radiologically. Light microscopically, the present case did not show the typical histology of conventional schwannoma and the confirmative diagnosis was made with the aid of immunohistochemistry and electron microscope. Here, we review the possible hypotheses for pathogenesis of sellar schwannomas on unusual locations.

Key Words : Sella Turcica-Neurilemmoma-Pituitary Neoplasms

Schwannomas are common benign tumors arising from cranial or spinal nerve roots, but only rarely do they occur in the intrasellar and parasellar regions.¹⁻⁶ We report a rare case of pituitary schwannoma that was initially thought to be a nonfunctioning pituitary macroadenoma. During surgery, a highly vascular tumor was adherent to the dura of the cavernous sinus, which was quite unusual for a pituitary adenoma. The pathologic diagnosis was a primary intrasellar schwannoma. The radiological and clinical aspects of the present case have been reported.⁷ Here, we focused on pathogenesis of sellar schwannomas.

CASE REPORT

The patient was a 39-year-old man with a sellar mass, which was incidentally found on computed tomography taken for evaluation of chronic sinusitis two months earlier. On checking his clinical history, the patient was found to have a history of bitemporal hemianopsia and slightly decreased libido for seven months. Sellar magnetic resonance images showed a gadolini-

um-enhanced mass, measuring 24 × 23 mm, in the widened sellar turcica (Fig. 1). The mass compressed the optic chiasm and slightly displaced the right cavernous sinus and carotid artery. But there were no evidences of tumor extension around the vessel or through the cavernous sinus. The mass displaced the infundibulum to the left side and extended cephalad. Radiologic diagnosis was pituitary macroadenoma. There were slight changes in the pituitary function test; the growth hormone (GH) level had slightly decreased to 0.59 ng/mL (normal: 1-5), adrenocortico tropic hormone (ACTH) was 4 pg/mL (normal: 0-60), and thyroid-stimulating hormone (TSH), prolactin, follicle-stimulating hormone (FSH), luteinizing hormone (LH), thyroxin (T4), triiodothyronine (T3) and testosterone levels were all within normal limits. The free T4 level was 0.72 ng/dL (normal: 0.89-1.8). Under the impression of pituitary macroadenoma, a transphenoidal approach was followed to the sella through a sublabial incision. A near total resection was performed and complete resolution of the visual field defect was found. During 30-month of postoperative follow-up, he was well without any evidence of recurrence or symptoms.

PATHOLOGICAL FINDING

The removed specimen was composed of aggregates of pinkish tan friable tissue and measured 2 cc in toto. The fresh specimen was fixed in 2.5% glutaraldehyde for electron microscopy and the remaining tissue was fixed in 10% neutral formalin. It was paraffin embedded and routine hematoxylin-eosin staining

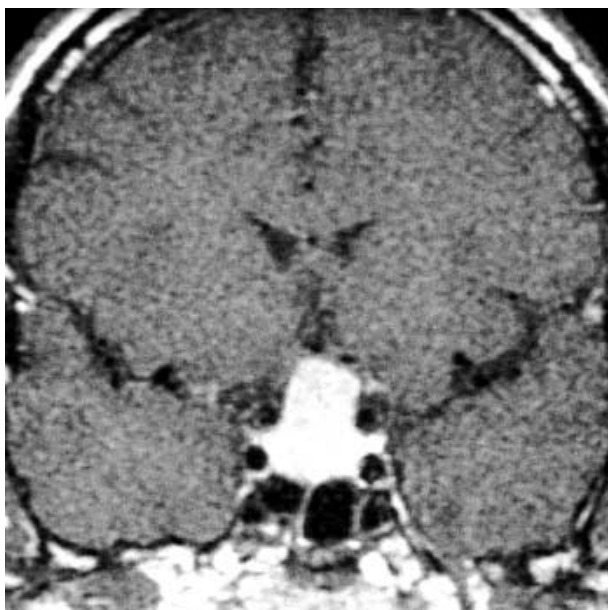


Fig. 1. T1-weighted coronal magnetic resonance image shows a gadolinium-enhancing mass in the sella turcica.

was carried out. For differential diagnosis from meningioma, glioma or pituitary adenoma, immunohistochemistry was performed using the antibodies (DAKO, Glostrup, Denmark) to S-100 protein (1:1,200 dilution), smooth muscle actin (1: 100), glial fibrillary acidic protein (1:350), epithelial membrane antigen (1:200) and pituitary hormones (GH 1:100, ACTH 1: 1,000, prolactin 1:800, LH 1:100, FSH 1:100, TSH 1:100). Light microscopically, the lesion was mainly composed of spindle cells arranged in small whorls and short fascicles (Fig. 2). The individual cells showed elongated and wavy cytoplasm with indistinct cell borders. The nuclei were oval with fine to hyperchromatic chromatin and indistinct small nucleoli. Among the spindle cells, slit like capillaries were scattered within the spindle cells. Focal microcystic changes, infiltration of inflammatory cells and extravasated red blood cells were also found. Neither mitosis nor necrosis was found. The tumor cells were diffusely strong immunoreactive for S-100 protein and were totally negative for smooth muscle actin, glial fibrillary acidic protein, epithelial membrane antigen or pituitary hormones. Under the light microscope, there were no perivascular hyalinization or Antoni A, B including Verocay bodies. The immunohistochemical properties of the tumor were suggestive of schwannoma. Ultrastructurally, the tumor cells were surrounded by duplicated continuous basal lamina (Fig. 3), giving a confirmative clue to the diagnosis of schwannoma.

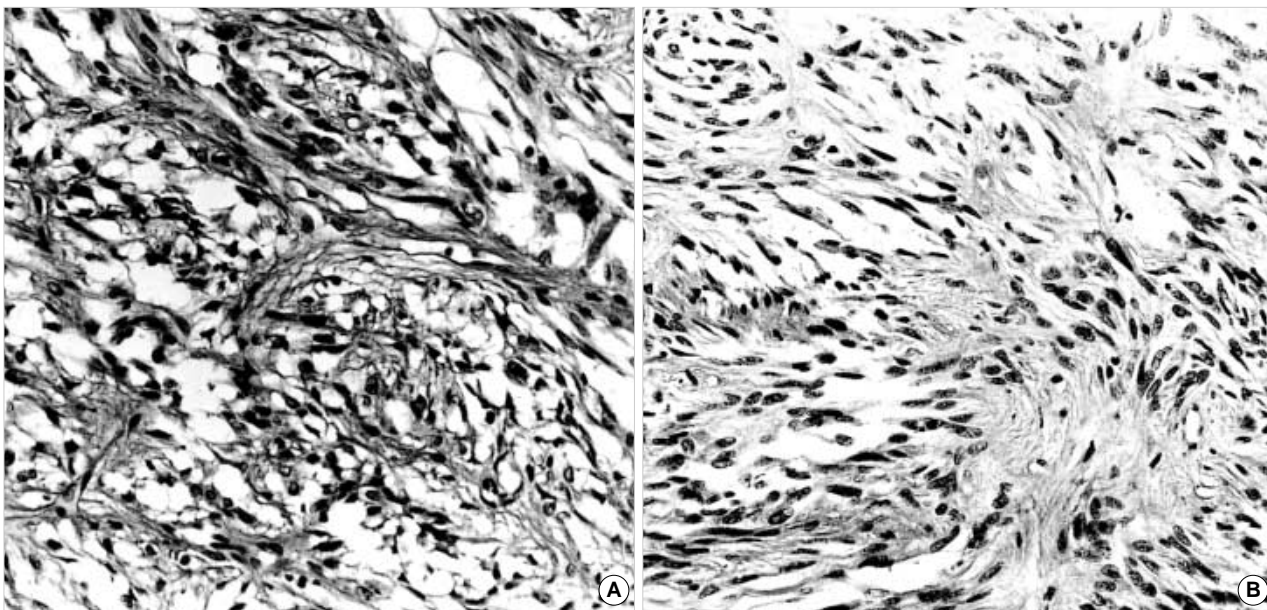


Fig. 2. (A) Histologically, the tumor is composed of loose bundles of the spindle cells with elongated nuclei and indistinct cytoplasm. (B) Focally, the spindle cells form an acellular collagenous area.

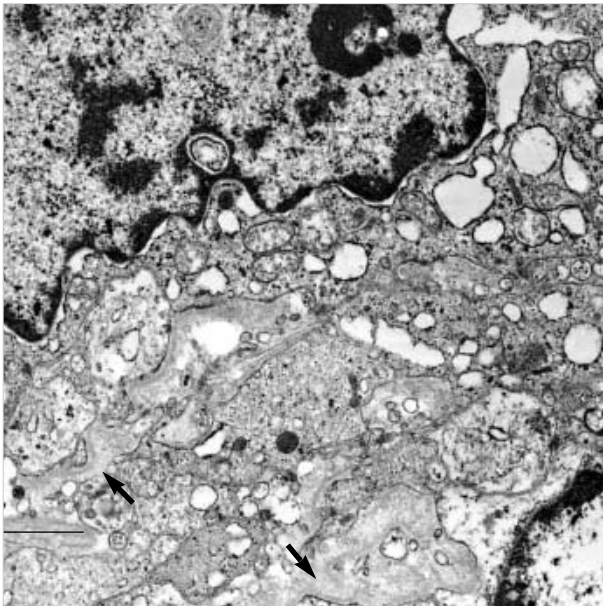


Fig. 3. Electron microscopic photograph of sellar schwannoma shows duplicated continuous basal lamina (arrows) surrounding tumor cells (original magnification $\times 6,000$, bar represents 10 nm).

DISCUSSION

Space occupying lesions of the sella turcica include pituitary adenoma, meningiomas, craniopharyngiomas, chordomas, germinomas and nonneoplastic cysts of inflammatory conditions and so on.⁸ Occasionally, parasellar schwannomas arising from oculomotor, trigeminal or trochlear cranial nerves or intracerebral schwannomas have been reported,^{3,9,10} and primary intrasellar schwannomas are more rare.¹⁻⁵

Histologically, most schwannomas are readily recognized by their distinctive alternation of dense fascicular Antoni A areas, spongy Antoni B tissue with small dark nuclei, and variably formed Verocay bodies. A collagenous capsule, hyalinized vessels, xanthoma cells, and perivascular hemosiderin complete the diag-

nostic picture. However, the present case showed a spindle cell tumor without the typical histologic features of schwannoma, rather resembling a fibrous meningioma or glioma. Moreover, this case occurred in an unusual location, which may have diagnostic problems, largely because the diagnosis of Schwann cell tumor does not come to mind. Once considered, the lesion is readily recognized on the basis of its immunohistochemical and electron microscopic features. Immunohistochemically, schwannomas demonstrate generalized strong staining for S-100 protein. Ultrastructurally, the elongated cells of schwannomas are individually surrounded by duplicated basal lamina. The present case showed both typical immunohistochemical and electron microscopic findings of schwannoma. Histologically, the present tumor simulated fibrous meningioma. However, schwannomas can be distinguished from meningiomas by the absence of both vague meningothelial whorls and membranous immunoreactive tumor cells for epithelial membrane antigen.¹¹ Meningioma may be reactive for the S-100 protein but it is often weak and patchy. Meningiomas also show a membranous pattern of immunoreaction for epithelial membrane antigen. The diagnostic problem from glioma was solved by the absence of glial fibrillary acidic protein-reactivity. Pituitary adenoma was easily excluded by not only the histology of the tumor but also the lack of immunoreactivity for pituitary hormones. Therefore, schwannomas arising in an unusual location such as this case may require immunohistochemical and ultrastructural application for the diagnosis.

On excluding schwannomas of the trigeminal, trochlear or oculomotor nerves with secondary involvement of the sella,¹²⁻¹⁴ five cases of primary sellar pituitary schwannomas have been reported in the literature.¹⁻⁵ The previously reported cases simulated pituitary tumors both clinically and radiographically. The presenting symptoms and suggested pathogeneses are briefly summarized in Table 1.

On the viewpoint of pathogenesis, Schwann cells are not natu-

Table 1. Literature review of primary intrasellar schwannomas

| Reference | Authors & Year | Age (yrs), Sex | Clinical Presentation | Possible Pathogenesis |
|-----------|---------------------|----------------|--|---|
| 1 | Goebel et al., 1979 | 25, F | Grand mal seizure | Schwann cells of sensory nerve or ectopic Schwann cells |
| 2 | Perone et al., 1984 | 39, M | Incidentally found after a minor head trauma | Perivascular Schwann cells |
| 3 | Wilberger, 1989 | 62, F | Visual loss for several years | Not mentioned |
| 4 | Guenot et al., 1994 | 67, F | Poor visual acuity Panhypopituitarism | Ectopic Schwann cells of perivascular area in the sella |
| 5 | Civit et al., 1997 | 41, M | Bitemporal hemianopsia | Perivascular Schwann cells or Schwann cells ensheathing the nerve twigs of the dura |

ral components of the pituitary gland and the origin of primary sellar schwannomas has been a matter of controversy. The cellular origin of primary intrasellar schwannomas remains unsettled. Several hypotheses have been advanced. There are two widely acceptable theories. First, the origin of sellar schwannomas can be explained on an embryonic basis, i.e. the possibility of ectopic or displaced Schwann cells resting within the sella.¹⁵ At the time of neural tube closure, neural crest cells divide in the midline, migrate laterally and become segmented into cell clusters between the neural tube and the somites.¹⁶ The neural crest cells migrate widely in the whole body and undergo various differentiations in different tissue, i.e. chromaffin cells, pigmented cells, autonomic and peripheral nervous system, sympathetic or sensory ganglia of cranial nerves. Based on this embryologic process, these Schwann cell nests pinched off in the segmentation of the neural crest are probably left in the intracerebral pituitary gland, leading to the source of sellar schwannomas. Second, perivascular Schwann cells in the pituitary gland are another possible source. The pituitary gland belongs to the endocrine organ in the cranial cavity and has a rich vascular supply. Therefore, certain stimuli evoking proliferation of perivascular Schwann cells in the pituitary gland may form schwannomas.¹⁷ There are two other possibilities; nerve twigs of the sensory or vasomotor nerves in the dura and pia covering the pituitary gland¹⁸ and conversion of pial cells into Schwann cells.¹⁹ Supporting evidences of those theories are as follows: the supplying nerve twigs covering the pituitary are branches of the trigeminal nerve, and histological similarity exists between the mesodermal cells of the pia and the neuroectodermal cells having derived from the neural crest. Despite the existence of several theories, none has yet been proved.

In conclusion, sellar schwannoma may show histologic findings different from conventional schwannoma in other sites. Sellar schwannoma might be included in the preoperative differential diagnosis of the sellar mass featuring only mass effect, although the incidence of intrasellar schwannoma is quite low in this location.

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