

Intrasellar Schwannomas

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The authors present two cases of intrasellar schwannoma, a condition rarely reported in the literature. The patients presented with symptoms of hypopituitarism. Sellar magnetic resonance imaging(MRI) revealed intrasellar masses extending into the suprasellar region, with the normal pituitary gland also identifiable in the imaging study. The tumors were removed via trans-sphenoidal route, and found to be very firm, unlike usual pituitary adenomas. Tissue diagnosis confirmed the tumors as schwannoma.

KEY WORDS : Schwannoma · Pituitary neoplasms · Sellar turcica · Hypopituitarism.

Introduction

Pathologic lesions involving the sellar or parasellar region include astrocytoma, chordoma, craniopharyngioma, dermoid cyst, germinoma, meningioma, and pituitary adenoma, with the latter being the most common pathology⁶⁾. Differential diagnosis is important in both predicting the prognosis and planning the treatment strategy. Intracranial schwannomas comprise six to eight percent of all primary intracranial neoplasms⁸⁾ and are most frequently located in the cerebello-pontine angle originating from the vestibular component of the eighth cranial nerve. Five reports of intrasellar schwannomas have previously been made^{1,4,7,9,10)}.

Case Report

Case 1

A 34-year old man presented with a history of headache over 5 months. Brain computed tomography (CT) revealed an intrasellar mass. On neurologic examination, the patient's visual acuity and field were normal. Physical examination revealed symptoms and signs of hypothyroidism, including bradycardia and decreased deep tendon reflex. Magnetic resonance imaging(MRI) showed a well-defined solid mass measuring 1.4 × 1.9 × 1.8cm in the sella and suprasella area. The signal of the lesion was iso-intense to cerebral cortex in T1-weighted image, and slightly hyper-intense in T2-weighted image. The mass was homogeneously enhanced after admini-

stration of Gd-DTPA. The mass partially compressed the optic chiasm, but did not invade the cavernous sinus (Fig. 1). Preoperative endocrine tests revealed hypothyroidism : TSH elevated to 17.02mIU/L (normal, 0.17-4.05mIU/L), T4 decreased to 3.28g/dl (normal, 4.2-12g/dl). Initial diagnosis was pituitary hyperplasia due to hypothyroidism, and the patient was given thyroid hormone for 6 months. Follow-up endocrine tests showed euthyroid state, but the mass size did not change. The patient then underwent trans-sphenoidal surgery for biopsy. When the dura was open, normal pituitary gland was noticed

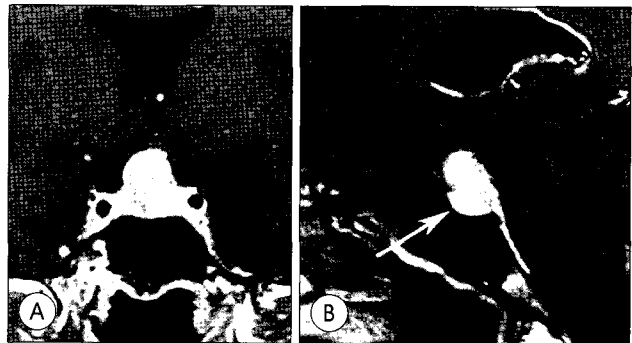


Fig. 1. Coronal (A) and sagittal (B), T1-weighted, gadolinium-enhanced, magnetic resonance images demonstrate an intrasellar mass partially compressing the optic chiasm and nerve. The lesion shows homogeneous enhancement and normal pituitary gland below the mass (arrow).

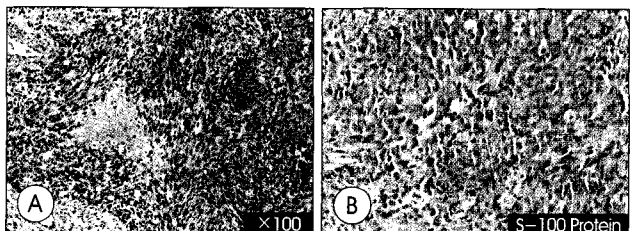


Fig. 2. The tumor exhibits a typical schwannoma pattern of spindle cells with palisading arrangement (H&E×100) (A) and is uniformly immunoreactive for S-100 (immunohistochemical stain×200) (B).

• Received : June 2, 2004 • Accepted : September 22, 2004
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on the sellar floor. The tumor was found to be firmer than usual pituitary adenoma and attached more tightly to the surrounding tissue. The mass was removed partially. Microscopic examination showed spindle cells with a palisade arrangement. Immunohistochemical staining for S-100 protein was positive, while that for EMA and GFAP was negative (Fig. 2).

Case 2

A 61-year old man presented with a history of headache and progressive visual dimness over 6 months. He also showed symptoms of polydipsia and polyuria. Neuro-ophthalmologic examination revealed bitemporal hemianopsia and decreased visual acuity. Endocrine evaluation showed partial pituitary-diabetes insipidus and elevated serum prolactin (168ng/ml). Other pituitary hormone levels decreased. MRI demonstrated a well-defined solid mass measuring 2.4×1.9×3cm in the sella and suprasella area. The mass was homogeneously enhanced after administration of gadolinium, but a suspicious normal pituitary gland was noticed below the mass. The optic chiasm was severely compressed by the mass, although there was no evidence of cavernous sinus invasion (Fig 3). We clinically diagnosed a non-functioning pituitary macroadenoma, and the mass was removed by trans-sphenoidal route. Sellar floor erosion was not prominent despite the macroadenoma. During the operation the mass was detected to have a hard consistency unlike usual pituitary adenoma. Abundant yellowish cheese-like material was also noted inside the mass. Normal pituitary gland was identified near the sellar floor and confirmed by frozen biopsy. Surgery sought total removal of the tumor. Following removal of the mass, the arachnoid matter that had been over the mass was displaced downward. Histopathologic examination showed cells with a palisade arrangement and typical Antony type A and B portions. Immunohistochemical staining for S-100 protein was positive, while that for EMA and GFAP was(Fig. 2).

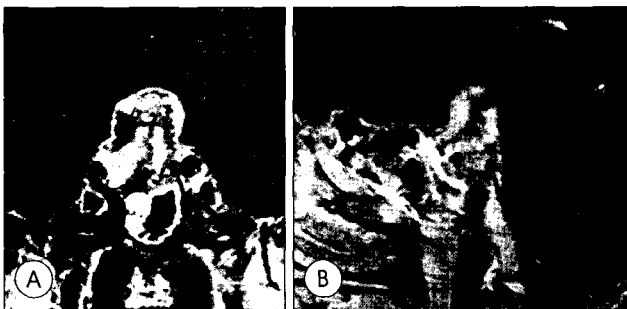


Fig. 3. Coronal (A) and sagittal (B), T1-weighted, gadolinium-enhanced, magnetic resonance images show an intrasellar mass expanding to suprasellar area. The lesion demonstrates heterogeneous enhancement after gadolinium infusion.

Discussion

The Schwann cells ensheath peripheral axons from the point at which they exit the pia mater to their terminations. Because the Schwann cells are not intrinsic components of the central nervous system, intra-axial schwannomas unrelated to cranial nerves are rare, and particularly so in the absence of neurofibromatosis Type 1. Four histopathological hypotheses have been put forth. Some reports of intrasellar schwannomas name the cranial nerves of the cavernous sinus, such as the oculomotor or trigeminal nerve, as the origin of the tumors^{3,7}. This is not appropriate to all cases, however. Russell and Rubinstein suggest that the ectopic Schwann cell is the origin of the tumors⁶. The third theory is that schwannomas with a tight attachment to the dura arise from Schwann cells ensheathing the small nerve twigs that innervate the dura⁸. Finally, other authors suggest that perivascular Schwann cells are the origin of the tumors^{2,5}. These reports confuse the issue of finding the tumors' origin, and the exact origins were not identified in the cases reported here. The masses were located at the midline and had no cavernous sinus involvement, so we concluded that the tumor origins were necessarily far from the cranial nerves of the cavernous sinus. Normal pituitary glands were found below the mass near the sellar floor, so we hypothesized that tumor growth started from the diaphragm sellar level before propagating upward and downward. Ectopic theory of tumor origin proposed by Russell and Rubinstein is the explanation most consistent with the present findings.

Differentiation of intrasellar schwannomas from pituitary adenomas is difficult. There are no unique MRI findings in intrasellar schwannomas which show iso-signal intensity on T1 weighted image and high-signal intensity on T2 weighted image. Enhancement patterns are various from homogenous to heterogenous. All the reported cases were diagnosed as pituitary adenoma initially, so the trans-sphenoidal approach had commonly been used. Some neurosurgeons, however, argue that the mass remains hard to manage and so maintain that the transcranial approach is appropriate whenever evidence of schwannoma exists^{7,10}. Here, we could accomplish partial removal in case 1 and total removal in case 2. Fortunately, the schwannoma, as an encapsulated benign tumor, did not require secondary operation in the former case. The operation findings are more similar to usual cerebello pontine angle schwannomas than pituitary adenomas. Visual symptoms and diabetes insipidus both improved following surgery in the latter case. There was no surgical morbidity in either case and tissue diagnosis was possible. We suggest that the approach indications of intrasellar schwannomas are not different from pituitary adenomas.

Immunohistochemistry and electron microscopy are needed for schwannoma diagnosis. To distinguish unusual intrasellar masses such as meningioma and schwannoma, EMA, S-100 protein, Vimentin, and GFAP are used. Although some limitations exist, schwannomas have positive reactions to S-100 protein and Vimentin, meningiomas to EMA, and pituitaryomas to GFAP⁸. Both of the cases reported here showed positive reactions to S-100 protein and Vimentin, so the masses were as intrasellar schwannomas.

Conclusion

Intrasellar schwannomas are a very rare form of intracranial schwannomas. Clinical presentations do not differ from usual pituitary adenoma, although surgical findings are unique. Authors report two cases of intrasellar schwannomas, as confirmed by tissue diagnosis.

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