



Review

Hard calcified intrasellar schwannoma mimicking pituitary adenoma: A case report and review of the literature



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ABSTRACT

Background: Intrasellar location of schwannoma is extremely rare, although intracranial schwannomas are common in the central nervous system. The aim of the present study is to describe a calcified intrasellar schwannoma case.

Materials and methods: We represent a 45-year-old woman who had suffered from headaches; right side facial pain and visual disturbance which had worsen during the last week prior to admission. Physical examinations were normal except for the bitemporal visual field hemianopia which match with perimetry examination. MRI demonstrated an unusual seemingly calcified mass lesion in the sellar region which was mimicking pituitary macro adenoma.

Result: Total resection of the tumor achieved through endoscopic transnasal transsphenoidal approach by extracapsular dissection and pathologic examination of the tumor revealed calcified schwannoma.

Conclusion: The differential diagnoses of sellar and suprasellar lesions include pituitary adenomas, craniopharyngiomas, meningiomas, and many others. However, schwannoma is not usually included, because the occurrence of schwannoma in the sellar or suprasellar region is extremely rare. Only few cases of intrasellar schwannomas have been reported in the literature, all of which presented a suprasellar extension similar to that of our case. Fascinating surgical point is managing very firm tumor through transsphenoidal corridor which we handle it by very sharp, debulking and extracapsular removal.

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1. Introduction

Schwannomas are relatively common tumors comprising 8–10% of all primary intracranial neoplasms; however the intrasellar location of schwannoma is extremely rare [1]. Schwannomas generally originate from the sensory nerves and have the potential to occur anywhere Schwann cells exist. This is while they have been recorded in virtually every site intracranially, including intracerebrally [2].

We present an unusual case of a calcified intrasellar schwannoma radiographically and clinically simulating a pituitary adenoma. The patient had been suffering from headaches, right side facial pain and visual disturbance.

Intracranial schwannomas commonly arises from the eighth cranial nerve in the cerebellopontine angle. Schwannoma arising from the sella and extending into the suprasellar region are very rare and are easily mistaken for pituitary adenoma.

2. Materials and methods

2.1. Case report

A 45-year-old woman came to us with headaches; right side facial pain and visual disturbance from 6 months ago which had worsened during the last week. Physical examinations were all normal except bitemporal visual field hemianopia which matches with perimetry examination. Laboratory findings and hormonal profile were normal. Magnetic resonance imaging (MRI) demonstrated a lesion in the sellar region which was isointense to hypointense on T1-weighted and hypointense on T2-weighted images extending to suprasellar cistern and compressing the optic chiasm. Following contrast injection there was an enhancement in the dome of lesion which represented compressed pituitary gland and another globular enhancement next to right carotid (Fig. 1).

Because of the presence of flow void like signal on coronal T1-weighted image near the carotid artery (although it also characterize calcification) we performed CT angiography to rule out vascular anomaly such as carotid aneurysm (Fig. 1 H and I). It showed normal vascular anatomy and separate adjacent dens calcification near the carotid artery.

Some contradictory imaging findings as presence of bulky calcification in the sellar region and unusual intrinsic intensity (such as hypointensity on T2-weighted MRI and unusual enhancement pattern) was harbinger of an unfamiliar pathology other than the everyday pituitary macroadenoma. Patient's right facial pain (constant burning pain in periorbit and cheek) also added to our concern about diagnosis.

We approached the lesion through transsphenoidal corridor. Assisted by our rhinologist colleague we exposed sellar floor endoscopically. After drilling the sellar floor and opening the dura we encountered grayish firm tumor (Fig. 2). Its consistency was rubbery that it could not be cut easily by knife. We decompress the lesion by punching it by otolaryngology throughcut punch. After internal debulking we proceed by extracapsular mean to dissect and resect the tumor. Its adherence to right cavernous sinus and carotid was significant. Superiorly diaphragm was opened and we tried to save compressed pituitary gland and stalk. By exploring the suprasellar cistern we removed the tumor totally and mammillary bodies and vascular structures showed up (Fig. 3). Repair of the sellar defect was done with autologous fat and fascia lata. A nasoseptal flap turned and covered the defect.

Histopathologic examination (Fig. 4) recommended schwannoma versus meningioma which immunohistochemistry (IHC) confirmed its diagnosis in favor of schwannoma which was highly positive for S100 antigen, positive for vimentin and negative for epithelial membrane antigen (EMA).

Postoperatively, the patient awoke without difficulty and was transferred to the intensive care unit for 24 h after surgery. There was no postop diabetes insipidus. Visual complaints abated satisfactorily. There was no complaint of facial pain anymore and she released from hospital on day three.

Postop course was uneventful. MRI 3 months after surgery showed total resection of the tumor, saved pituitary gland and small fat inside the sella which we used for prevention of CSF leak (Fig. 5). Hormonal profile was normal after surgery and the patient did not need any hormonal replacement therapy.

Because of her preoperative facial pain and tumor origin from the right cavernous sinus and finding schwannoma on histopathology it would be reasonable to postulate the tumor was a schwannoma of ophthalmic or maxillary branch of trigeminal nerve, although we cannot prove that. After surgery no extraocular muscle problem occurred and corneal sensation and facial senses was intact.

3. Discussion

Most neoplasms growing in the sella turcica are pituitary adenomas. The remaining neoplasms are benign tumors (craniopharyngiomas and meningiomas), malignant tumors (primary germ cell tumors, chordomas, lymphomas, and pituitary adenocarcinomas, or metastatic disease mainly from breast and lung cancer), and other uncommon conditions (cysts, abscesses, arteriovenous fistulas, lymphocytic hypophysitis) [3]. As mentioned before, schwannoma accounts for up to 8% of primary brain tumors [4], and is usually associated with the cranial nerves and shows a special predilection for the vestibular portion of the 8th cranial nerve at the cerebellopontine angle, while intrasellar localization is exceptional.

The most common differential diagnoses of sellar and suprasellar lesions include pituitary adenomas, craniopharyngiomas, and meningiomas; however, schwannoma is not usually included. Only few cases of intrasellar schwannomas have been reported previously (Table 1), all of which presented a suprasellar extension similar to that of our case [5].

The clinical and radiological presentation of intrasellar schwannomas is consistent with the findings of pituitary adenomas. The most common signs and symptoms of intracranial schwannomas are headaches, seizures, and focal neurologic deficits [1]; similar to our case which was developed with headaches, and bitemporal hemianopia. Other pathologic characteristic and imaging features of intraparenchymal schwannomas include calcification, cyst formation, peritumoral edema and/or gliosis, and superficial or periventricular location [1]. As expected, the MRI findings of this study showed that the lesion has cystic component in the sellar region and has been calcified [2].

In a study conducted by Whee et al. it was revealed that the enlargement of the sella turcica and an enhancing sellar mass with suprasellar extension are most often associated with pituitary adenomas [5] and radiological findings in these cases were not significantly different from the usual pituitary macroadenomas. In an excellent review on the unusual lesions in the region of sella, Chaddock [6] reported one patient with a trigeminal schwannoma secondarily involving the suprasellar region. Afterwards, Goebel et al. [7] described a schwannoma reportedly arising from tuberculum sella without associated extension into the sella.

As there is no obvious nerve within the sella, the origin of primary intrasellar schwannomas supposed to be from lateral sellar nerve plexus, perivascular schwann cells, or sensory nerves of the dura [8–10]. Firstly, lateral sellar nerve plexus is a distribution center for visceromotor and sensory nerves, which innervate cerebral

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