

# Intrasellar schwannoma mimicking pituitary adenoma

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## ABSTRACT

Intrasellar location of schwannoma is extremely uncommon and 18 cases are documented in the literature till now. This report describes intrasellar schwannoma in a patient in whom the neuroimaging features were suggestive of a pituitary adenoma.

**Key words:** Intrasellar schwannoma, neuroimaging, pituitary adenoma

## INTRODUCTION

Intracranial schwann cell tumors account for 8% of all primary intracranial neoplasms.<sup>[1]</sup> Most frequently intracranial schwann cell neoplasms are located in the cerebello-pontine angle originating from the vestibular branch of the acoustic nerve. Intracranial schwannoma not related to cranial nerves are extremely rare, more so in the intrasellar location. Till date, 18 cases have been documented in the literature.<sup>[2]</sup> In this report, we describe a case of intrasellar schwannoma in a patient who presented with the clinical and neuroimaging features of a pituitary adenoma.

## CASE REPORT

This patient, a 67-year-old female, a known hypertensive, presented to us with features of hypopituitarism. Investigations revealed a sellar mass with supra sellar extension. The lesion was well demarcated measuring 2.3 cm × 2.1 cm × 1.7 cm in dimensions. The lesion was isointense in both T1- and T2-weighted imaging. It was uniformly enhancing on contrast administration with lobulated borders [Figure 1]. The superior convex border of the mass was abutting the optic chiasm. There was no parasellar or retrosellar extension. The lesion was abutting the cavernous and supraclinoid carotid arteries on either side without significant encasement.

There was no orbital or sphenoid sinus extension. Normal T1-hyperintensity of posterior pituitary was not seen separately. Pituitary stalk was not well identified. Clinical examination revealed that she had bitemporal hemianopia and hypopituitarism. Hypopituitarism was corrected with hormonal supplementation and she was operated with a diagnosis of pituitary macroadenoma. A trans septal, trans sphenoidal microsurgical excision of the lesion was done. Patient had an uneventful post-operative recovery and had been coming for a regular follow-up. She is still hypocortisolemic and is on replacement therapy with oral prednisolone.

## Histopathology

H and E stained paraffin sections showed classical features of a schwannoma. The neoplastic cells were made up of spindle cells and they were displayed in a regular rhythmic palisading pattern (Antoni-A). At other microscopic fields loosely textured stellate cells (Antoni-B) were also present [Figure 2]. Large portions of the tumor consisted of acellular dense collagen. Nuclear atypical changes and mitotic activity were not demonstrated in the sections. There was no demonstrable evidence of epithelial component or neoplastic cells of pituitary origin. Immunostaining demonstrated that the tumor cells had intense expression of nuclear S 100 protein, without any expression of epithelial membrane antigen. The Ki-67 nuclear labeling index was approximately 1% indicating a slow proliferative potential. These above features were diagnostic of a schwannoma.

## DISCUSSION

About 9% of sellar tumors do not arise from adenohypophyseal cells<sup>[3]</sup> and these include primary sellar lesions, (craniopharyngioma, meningioma, schwannoma, chordoma) and metastatic tumors (breast, prostate,

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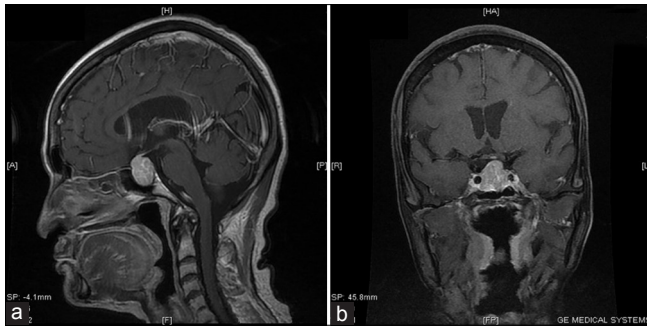
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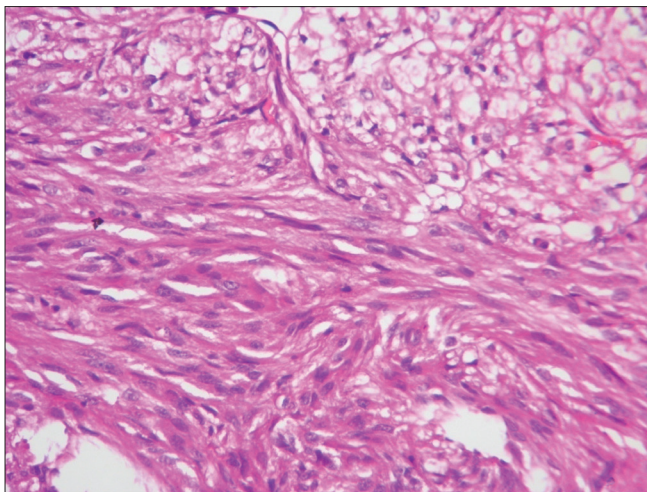
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**Figure 1:** Gadolinium enhanced T1-weighted magnetic resonance imaging images in sagittal and coronal planes showing the sellar mass with suprasellar extension



**Figure 2:** Photomicrograph showing classical Antoni A and B components of a schwannoma (Hematoxylin and eosin x400)

bronchus, renal cell carcinoma). In addition to these inflammatory lesions such as tuberculosis and sarcoidosis may also mimic a sellar mass lesion in magnetic resonance imaging (MRI) scans.<sup>[4]</sup> Among these non-pituitary tumors of the sella, schwannoma is the least common. A review of 18 published cases of sellar schwannoma revealed that they tend to occur in the fifth and sixth decade of life. In 14 cases, there was no demonstrable attachment to the cranial nerves. In one case tumor was totally demarcated from the pituitary structures.

The imaging features in MRI scans in intrasellar schwannoma include enhancement of sella turcica with an enhancing sellar mass with suprasellar extension. These imaging features are not different from that of a pituitary macroadenoma. Hence it will be impossible to diagnose an intrasellar schwannoma by imaging alone. It is therefore not surprising to note that in none of the 18 cases a pre-operative diagnosis of schwannoma could be made. The clinical presentation of intrasellar schwannoma depends largely on its size and impact on pituitary functions. The usual clinical features in these

patients are visual field defects and decreased visual acuity. Evidence of hypopituitarism is documented in 50% cases of intrasellar schwannoma. In one case, there was an intrasellar schwannoma with a co-existing growth hormone secreting pituitary adenoma.<sup>[5]</sup> Sellar region schwannomas are typically well circumscribed and vascular and firm in consistency. The marked vascularity of schwannomas increases the likelihood of intratumoral bleeding. The histopathological features of intrasellar schwannoma are quite similar to that of schwannomas arising from the vestibular nerve and hence do not present any diagnostic difficulties. Other spindle cell neoplasm of the sella such as spindle cell oncocytoma needs to be distinguished from schwannoma. Schwannoma can be distinguished from spindle cell oncocytoma of the pituitary by the combination of histopathological features as well as by positive immunostaining for S-100 in schwann cells.

As there are no nerves arising within the sella turcica, the origin of intrasellar schwannoma remains speculative. Several theories concerning the histogenesis and development of an intrasellar schwannoma has been proposed. According to Bleys *et al.*,<sup>[6]</sup> the human cavernous sinus contains an extensive nerve plexus with small ganglia and has connection with cranial nerves. The connection between lateral sellar plexus and functionally defined nerve structures would suggest that the intrasellar schwannoma could originate from the lateral sellar plexus. The perivascular nerve plexus in which schwann cells can be found is another theory for the origin of intrasellar schwannoma.<sup>[7]</sup> Other theories include the ectopic schwann cells due to the transformation of pial cells and pluripotential mesenchymal cells giving rise to schwannoma.<sup>[8]</sup> However, none of these hypotheses have been proven, so the exact histogenetic origin of an intrasellar schwannoma remains unclear.

Management of patients with intrasellar schwannomas should include evaluation of pituitary function, a neuro-ophthalmologic evaluation and a neuroradiological examination with MRI. Neurosurgical intervention and excision can be attempted either by trans-sphenoidal route or by trans cranial approach. By and large total excision of the lesion could not be achieved in intrasellar schwannoma. Though none of the reported patients required second operation for the remnant tumor, additional treatment such as radiosurgery needs to be considered because the follow-up period was too short to determine the prognosis.

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