Computed tomography and magnetic resonance imaging findings of intrasellar schwannoma: A case report and literature review

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Abstract. Intrasellar schwannoma is extremely rare, and only a few cases have been reported in the literature to date. In the present study, the clinical, radiological and pathological features of a 50-year-old male patient with primary intrasellar schwannoma are presented. The patient presented with decreased visual acuity, and a subsequent computed tomography (CT) scan revealed a giant well-demarcated intrasellar mass with suprasellar extension, without cavernous sinus invasion. The lesion was slightly hyperdense with heterogeneous enhancement. On magnetic resonance imaging (MRI), the mass was isointense on T1 weighted images (WI), and slightly hyperintense on T2WI, with significant heterogeneous enhancement. Previous literature was reviewed to summarize the CT and MRI characteristics of intrasellar schwannoma. Despite the rarity of this tumor, intrasellar schwannoma must be included in the differential diagnosis of intrasellar lesions.

Introduction

Schwannomas are common intracranial tumors, accounting for ~8% of all primary brain neoplasms (1). These tumors are benign and slow growing neoplasms of Schwann cell origin (1), which mostly affect middle aged (52.4±15.7 years) individuals (2). The overall 5-year survival rate is 79.6%, while the post-operative mortality is 0.5% (2). The majority of tumors originate from the vestibular portion of the auditory nerve at the cerebellopontine angle (3), while tumors arising from the trigeminal nerve are less common. Intrasellar schwannomas not associated with cranial nerves are extremely rare, and, to the best of our knowledge, only 17 cases have been reported in the English literature to date (3-17). Intrasellar schwannomas typically affect middle-aged and elderly individuals ranging from 33-79 years of age, with no significant gender preferences (3-17). Surgical resection is currently the leading treatment, and the majority of patients have a good prognosis. Furthermore, the majority of previous reports regarding intrasellar schwannomas have focused on the surgical approaches and immunohistochemical features of the tumor (4-7,9-12), whereas few studies have reported the computed tomography (CT) and magnetic resonance imaging (MRI) features of intrasellar schwannomas in detail. In the present study, a case of pathologically confirmed intrasellar schwannoma is presented, and the available literature is reviewed to summarize the radiological features of this lesion.

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Case report

In April 2014, a 50 year-old man presented to The First Affiliated Hospital of Chongqing Medical University (Chongqing, China) with decreased visual acuity that had lasted for 5 years, and had been aggravated for 1 week. Visual field examination did not identify visual field loss. The patient had a 10-year history of hypertension and a 2-year history of diabetes mellitus, and thus had been taking valsartan and metformin regularly; this medication was continued throughout the perioperative period. No abnormal findings were identified on complete physical and neurological examinations, which included assessments of the vital signs, muscle strength, muscle tone, sensory system and reflex system, in addition to cranial nerve examination. Preoperative endocrinological tests revealed normal levels of triiodothyronine (T3; patient level, 2.17 pg/ml; normal range, 2.19-3.90 pg/ml), thyroxine (T4; patient level, 0.91 ng/dl; normal range, 0.61-1.12 ng/dl), thyroid-stimulating hormone (TSH; patient level, 1.63 μIU/ml; normal range, 0.35-3.50 μIU/ml), adrenocorticotropic hormone (ACTH; patient level, 20.97 pg/ml; normal range, 7.20-63.30 pg/ml), prolactin (PRL; patient level, 6.55 ng/ml; normal range, 2.64-13.13 ng/ml), follicle-stimulating hormone (FSH; patient level, 21.72 mIU/ml; normal range, 1.27-19.26 mIU/ml), luteinizing hormone (LH; patient level, 2.65 mIU/ml; normal range, 1.24-8.62 mIU/ml), growth hormone (GH; patient level, 0.03 ng/ml; normal range, 0.00-5.00 ng/ml), cortisol (patient level, 169.93 nmol/l; normal range, 118.64-618.02 nmol/l) and testosterone (patient level, 1.80 ng/ml; normal range, 1.75-7.81 ng/ml).

A CT scan (Somatom Definition FLASH CT scanner; Siemens AG, Munich, Germany) revealed a 3.9x3.8 cm well-demarcated intrasellar mass with suprasellar extension, without cavernous sinus invasion (Fig. 1). The lesion was slightly hyperdense compared to the brain parenchyma,

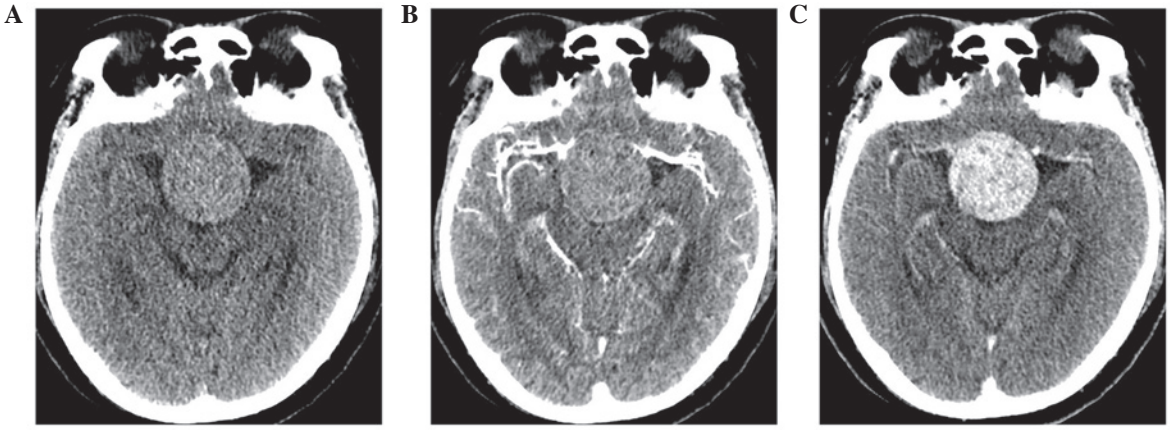

Figure 1. CT findings of intrasellar schwannoma. (A) Plain axial CT scan revealing a slightly hyperdense mass of the intra-suprasellar region. Following the administration of contrast agent, the mass shows heterogeneous enhancement at the (B) arterial phase and (C) venous phase. CT, computed tomography.

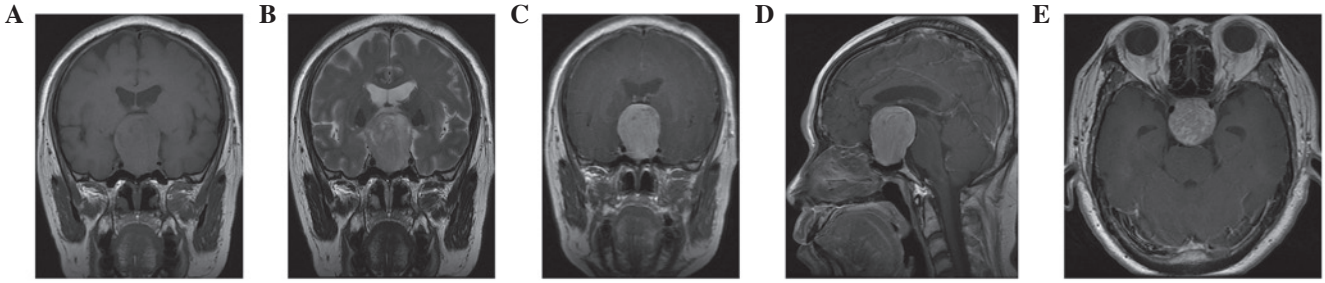

Figure 2. Magnetic resonance imaging findings of intrasellar schwannoma. The tumor appears as an (A) isointense T1WI and (B) slightly hyperintense T2WI mass, with remarkable heterogeneous enhancement on the (C) coronal, (D) sagittal and (E) axial planes. WI, weighted image.

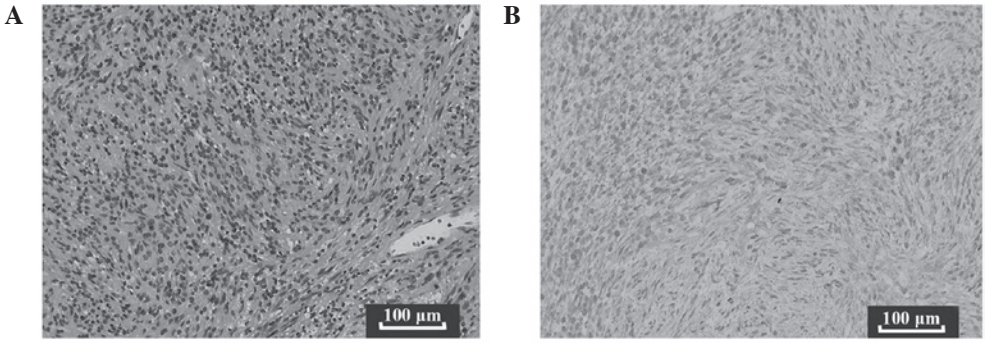

Figure 3. Pathological findings of intrasellar schwannoma. Microscopic examination demonstrated a tumor composed of (A) cells in a spindle arrangement, which are (B) strongly positive for S100. Staining, hematoxylin and eosin (Bogoo Biomart, Shanghai, China); magnification, x400.

with heterogeneous enhancement. On MRI (Signa HDxt 3.0T scanner; GE Healthcare Bio-Sciences, Pittsburgh, PA, USA), the lesion was isointense on T1 weighted images (WI), and slightly hyperintense on T2WI images, with significant heterogeneous enhancement following the intravenous administration of gadolinium diethylenetriamine pentaacetic acid (Bayer AG, Leverkusen, Germany) (Fig. 2).

Non-functioning pituitary adenoma was diagnosed preoperatively. Subsequently, the patient underwent gross total resection via an endonasal transsphenoidal approach. The resected mass was soft, grayish and hypervascular. Microscopic examination (using a XSG-111L microscope; Shunyu, Zhejiang, China) revealed a tumor composed of cells arranged in a spindle (Fig. 3). Immunocytochemistry was performed on the tumor cells by the Pathology Department at The First Affiliated Hospital of Chongqing Medical University (Chongqing, China), which revealed positivity for S100, and negativity for glial fibrillary acidic protein and epithelial membrane antigen. Consequently, schwannoma was diagnosed. Postoperative endocrinological tests revealed panhypopituitarism. The results of the tests were as follows: T3, 1.79 pg/ml (normal range, 2.19-3.90 pg/ml), T4, 0.90 ng/dl (normal range, 0.61-1.12 ng/dl), TSH, 0.12 μIU/ml (normal range, 0.35-3.50 μIU/ml), ACTH, 6.86 pg/ml (normal range, 7.20-63.30 pg/ml), PRL, 3.27 ng/ml (normal range, 2.64-13.13 ng/ml), FSH, 8.43 mIU/ml (normal

Table I. Intrasellar schwannomas: Literature review.

First author, year	Case, no.	Gender	Age, years	Clinical symptoms	Endocrine status	CT findings		MRI findings			(Ref.)
						Density	Enhancement	T1	T2	Enhancement	
Perone, 1984	1	M	39	Headache for 6 years	Normal	Iso	Homo	NA	NA	NA	(17)
Wilberger, 1989	2	F	62	Visual loss for several years	Panhypopituitarism	NA	Homo	NA	NA	NA	(16)
Civit, 1997	3	M	41	Bitemporal hemianopsia	Normal	NA	NA	NA	NA	Homo	(15)
Bhagat, 2002	4	M	51	Erectile impotence, fatigue and lethargy for 5 years	Panhypopituitarism	NA	NA	NA	NA	Homo	(14)
Bhagat, 2002	5	M	68	Visual loss for 2 years	Panhypopituitarism	NA	Homo	NA	NA	NA	(14)
Whee, 2002	6	M	39	Decreased visual acuity for 10 months	Panhypopituitarism	NA	NA	Iso	↑	Homo	(13)
Maartens, 2003	7	F	33	Headache for 6 months, decreased visual acuity, amenorrhea	Elevated prolactin, hypothyroidism	NA	NA	NA	NA	Hetero	(12)
Maartens, 2003	8	F	56	Bitemporal hemianopia, ataxia	Hypopituitarism	NA	NA	NA	NA	NA	(12)
Esposito, 2004	9	M	73	Lipothymia, bitemporal hemianopia	Panhypopituitarism	NA	NA	Iso	NA	Homo	(11)
Perez, 2004	10	F	71	Bitemporal quadrantanopia for 2 months	Normal	NA	Homo	Iso	NA	Homo	(10)
Honegger, 2005	11	F	79	Syncope, headache for 6 months	Hypothyroidism	NA	NA	NA	NA	Homo	(9)
Moreland, 2006	12	M	41	Headache for 3 years	Normal	NA	NA	NA	NA	Homo	(8)
Krayenbühl, 2007	13	M	NA	Headache, left superior temporal quadrantanopia	Panhypopituitarism	NA	NA	NA	NA	Hetero	(7)
Koutourousiou, 2009[a]	14	NA	38	Acromegaly	Elevated GH	NA	NA	NA	NA	NA	(6)
Park, 2009	15	F	49	Headache, vomiting and bitemporal hemianopsia	Normal	NA	NA	Iso	Slightly↑	Homo	(5)
Mohammed, 2010	16	F	45	Headache for 2 years, facial pain	Elevated prolactin, decreased FSH and LH	NA	NA	NA	NA	Homo	(4)
Cugati, 2012	17	M	42	Headache and visual loss for 1 year	Normal	Iso	NA	Iso	Slightly↑	Hetero	(3)
Present study	18	M	50	Decreased visual acuity for 5 years	Normal	Slightly↑	Hetero	Iso	Slightly↑	Hetero	-

[a]Represents a case of an intrasellar schwannoma co-existing with a GH-secreting pituitary adenoma. In consequence, the clinical presentations and endocrine changes are due to the pituitary adenoma rather than the schwannoma. CT, computed tomography; MRI, magnetic resonance imaging; M, male; F, female; NA, not available; ↑, hyperdensity or hyperintensity; iso, isointensity; homo, homogeneous; hetero, heterogeneous; GH, growth hormone; FSH, follicle-stimulating hormone; LH, luteinizing hormone; ref., reference.

range, 1.27-19.26 mIU/ml), LH, 2.53 mIU/ml (normal range, 1.24-8.62 mIU/ml), GH, 0.12 ng/ml (normal range, 0.00-5.00 ng/ml), cortisol, 265.13 nmol/l (normal range, 118.64-618.02 nmol/l) and testosterone, 0.36 ng/ml (normal range, 1.75-7.81 ng/ml).

The patient underwent replacement therapy with hydrocortisone (30 mg once per day) and Euthyrox (25 μg once per day, which were administered orally. MRI performed on follow-up examination 3 months following surgery revealed no evidence of tumor recurrence. Further MRI was planned at 6 month and 1 year follow-ups. Written informed consent was obtained from the patient.

To identify relevant studies, database searches of all literature published prior to November 26, 2014 on PubMed (Medline; www.ncbi.nlm.nih.gov/pubmed/) and Web of Science (www.webofknowledge.com/) were performed, using the search terms 'pituitary fossa', 'intrasellar', 'sellar' and 'schwannoma'. All searches were limited to English language publications. Suprasellar and parasellar schwannomas that did not affect the intrasellar regions were excluded. A manual search of the references of the retrieved articles was conducted subsequently. The search yielded 15 articles corresponding to 17 cases, in addition to the present case, which are all presented in Table I.

Discussion

Pituitary adenomas account for the majority of intrasellar tumors (8). The most common non-pituitary-derived tumors of the sella tunica include craniopharyngioma, glioma, meningioma and chordoma (4). As intrasellar schwannomas are extremely rare (4), they are not usually considered in the differential diagnosis of sellar tumors.

To date, only 18 cases of intrasellar schwannomas, including the present case, comprising of 10 males and 7 females (the gender of one case is not reported), with an average age of 51.6 years (range, 33-79 years) have been reported in the literature (3-17). In 12 of the 18 cases (66.7%), patients reported visual changes (particularly bitemporal hemianopsia), and 8 of the 18 cases (44.4%) suffered from headache. Less common clinical presentations included personality changes, erectile impotence, amenorrhea, syncope, vomiting and facial pain.

Only 3 cases (3,17) described plain CT scan features of intrasellar schwannomas, 2 of which appeared as an isodense mass, while the mass in the present case was slightly hyperdense. A total of 4 reported cases (10,14,15,17) exhibited enhanced CT features, all of which were reported to be homogeneously enhanced. However, the mass in the present case exhibited slight heterogeneous enhancement. Notably, marked cystic changes, intratumoral hemorrhage, calcification and necrosis were not identified in any of the 18 cases. All the 13 cases (3-5,7-15) that reported MRI features presented an isointense signal on T1WI and slightly hyperintense signal on T2WI, and the majority of these cases (9/13; 69.2%) showed homogeneous enhancement. In addition, the majority of cases exhibited a well-demarcated mass without cavernous sinus invasion. By contrast, a large number of pituitary adenomas invade the cavernous sinus (18). However, intrasellar schwannomas cannot be distinguished from pituitary adenomas based on such features alone (13).

In the present case, no specific nerve of origin was identified, similarly to the majority of reported cases, and thus, the origin of the intrasellar schwannoma remains unclear. However, three histopathogenetic hypotheses exist regarding the origin of intrasellar schwannoma. Firstly, Bleys *et al* (19) reported the existence of the lateral sellar nerve plexus within the cavernous sinus, which is a distribution center for visceromotor and sensory nerves innervating cerebral arteries, orbital structures and the dura mater. Notably, Dietemann *et al* (20) have revealed that there is not a clear separation between the pituitary fossa and the cavernous sinus, so there is a potential deficiency in the medial wall of the cavernous sinus. Certain authors (4,12) propose that tumors originating from the lateral sellar nerve plexus may extend through the the medial wall of the cavernous sinus to form an intrasellar schwannoma. However, the majority of reported cases do not involve the cavernous sinus (4-9). Secondly, it has been suggested that perivascular Schwann cells may be the source of intrasellar schwannomas (13). Small cerebral arteries of 10-15 μm in diameter are known to be accompanied by a perivascular nerve plexus, where Schwann cells may be located (21). Notably, intracerebral schwannomas originating from the perivascular nerve plexus have been reported (22,23). Therefore, it may be hypothesized that intrasellar schwannomas may originate from the perivascular nerve plexus of the inferior hypophyseal artery. Thirdly, Russell and Rubinstein (1) have suggested that intracerebral schwannomas may arise from ectopic Schwann cells; therefore, this theory may also explain the origin of intrasellar schwannomas.

In conclusion, intrasellar schwannomas are rare sellar tumors, which are commonly misdiagnosed as pituitary adenomas (13). Intrasellar schwannomas usually affect middle-aged and elderly individuals ranging from 33-79 years of age, with no significant gender preferences (3). The most common clinical presentations are visual changes and headache (3). On CT images, the majority of cases appear as a well-demarcated isodense intrasellar mass with homogeneous enhancement, without marked cystic change, intratumoral hemorrhage, calcification or necrosis (3,10,14,16,17). The majority of cases appear isointense on T1WI and slightly hyperintense on T2WI, with homogeneous or heterogeneous enhancement (3-5,7-15). The involvement of the cavernous sinus in intrasellar schwannomas is less frequent than that observed in pituitary adenomas (4,5,8,18). However, intrasellar schwannomas are usually indistinguishable from pituitary adenomas. Therefore, the final diagnosis is dependent on histology. Therefore, schwannomas should be included in the differential diagnosis of an intrasellar lesion.

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