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Case Report

Schwannoma in sellar region: report of two cases and a literature review

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Abstract: Background: Intracranial schwannomas are common primary brain tumors, which typically originate from the trigeminal nerve or the vestibular nerve. Schwannoma arising in the sellar region is an extremely rare clinical entity, which mimics pituitary adenoma and poses a great diagnostic challenge. Case description: Herein, we describe two cases of sellar schwannoma. In the first case, a sellar lesion was incidentally found in a 51-year-old asymptomatic female; surgical resection via the Trans-sphenoidal approach was performed. The postoperative course was uneventful. The second patient was a 50-year-old man who presented with a history of headache for 4 months. Magnetic resonance imaging showed a sellar lesion extending into the right cavernous sinus region. Surgical resection was performed via the right frontotemporal approach. Additionally, we reviewed the published literature and identified a total of 30 cases of sellar schwannoma. A brief analysis and a summary of the individual clinical manifestations, radiological characteristics, pathological findings, treatment details, and outcomes of these cases are presented. Conclusion: Intrasellar schwannoma is an exceedingly rare entity. Surgical resection is an effective treatment option. Preoperative differential diagnosis is typically challenging. Definitive diagnosis depends on histopathological and immunohistochemical evidence.

Keywords: Schwannoma, sellar region, surgical resection, case report

Introduction

Intracranial schwannomas are common entities, which account for approximately 8%~10% of all primary brain tumors [1]. These tumors are commonly located in the cerebello-pontine angle and originate from the trigeminal or the vestibular nerves [2]. However, intrasellar schwannoma is an extremely rare clinical entity. It typically mimics pituitary adenoma, thereby posing a diagnostic challenge [2]. The current literature regarding sellar schwannoma is primarily limited to case reports and does not provide a comprehensive overview. Thus, the clinical and radiological features as well as treatment and prognosis of sellar schwannoma are not well characterized.

Herein, we report two cases of sellar schwannoma. Additionally, we performed a literature re-view and identified a total of 30 relevant case reports. A brief analysis and a summary of the individual clinical manifestations, radiological characteristics, pathological findings, treatment details and outcomes are presented.

Case report

Case 1

The patient was a 51-year-old woman in whom a solid lesion in the sellar region was incidentally detected by magnetic resonance imaging (MRI) 15 days prior to admission. There were no notable abnormal signs or symptoms. General physical examination revealed no abnormality. On ophthalmologic examination, visual acuity in the right and left eyes was found to be 20/80 and 20/66, respectively. The intraocular pressure in the right and left eyes was 11 mmHg and 14 mmHg, respectively. Visual field examination revealed a scotoma in the left-upper-temporal quadrant. Neurological examination showed normal sensorimotor function and nervous reflexes in the extremities. Pituitary hormone levels were within the normal reference range. Brain MRI showed a 17 mm×14 mm×13

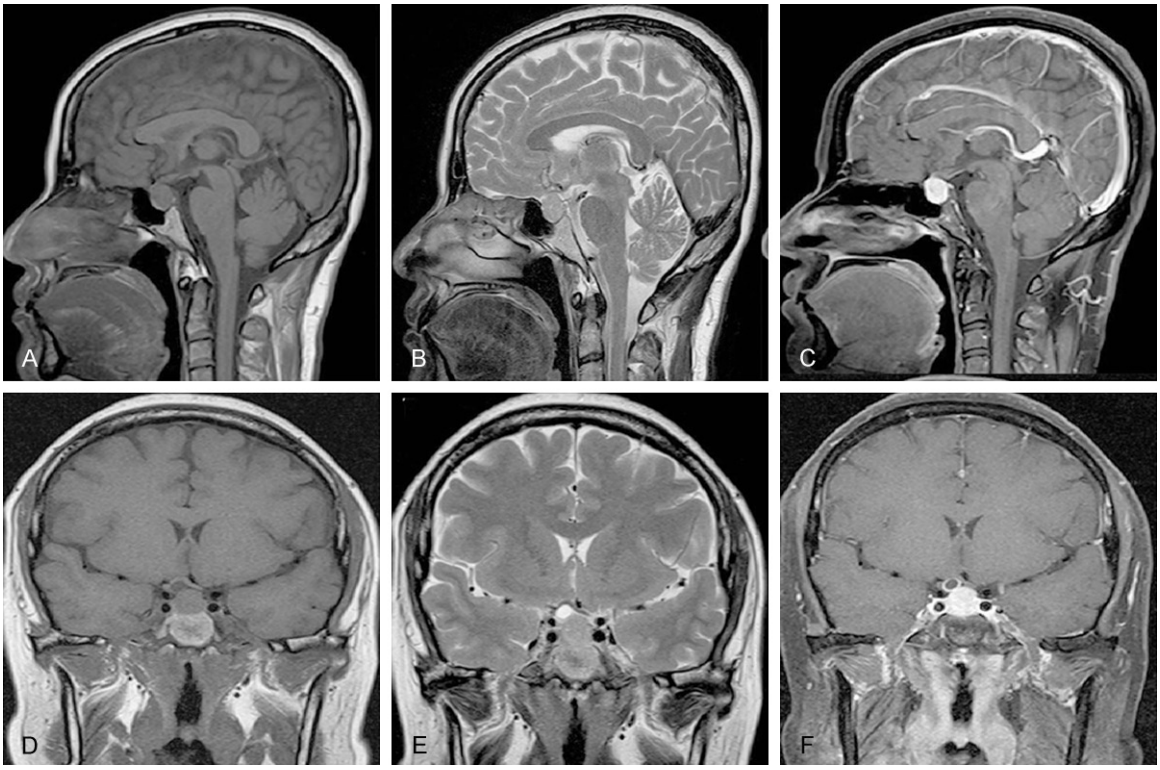

Figure 1. Magnetic resonance imaging of case 1. Sagittal (A) and coronal (D) T1-weighted images showing an isointense lesion in the sellar region. Sagittal (B) and coronal (E) T2-weighted images showing a slightly hyperintense lesion. Sagittal (C) and coronal (F) T1-weighted contrast-enhanced images showing remarkable homogeneous enhancement of the lesion.

mm oval lesion in the sellar region. The lesion appeared isointense on T1-weighted imaging and hyperintense on T2-weighted imaging and fluid attenuated inversion recovery (FLAIR) imaging. After administration of gadolinium-diethylene triamine pentaacetic acid (Gd-DTPA), the lesion showed homogeneous enhancement (**Figure 1**). Preliminary preoperative diagnosis was pituitary adenoma, and a surgical resection via the Trans-sphenoidal approach was scheduled. Intraoperatively, the tumor was confirmed to be located in the sellar region, and there were no significant adhesions with the optic chiasma. Histopathological examination revealed a cellular schwannoma, corresponding to World Health Organization (WHO) grade I. The postoperative course was uneventful; serial endocrine examination showed normal levels of pituitary hormones. As of 3-month follow-up, the patient remained asymptomatic.

Case 2

A 50-year-old male presented with a history of headache since 4 months, which relieved spontaneously after rest. There were no complaints of dizziness, nausea, or vomiting. General physical examination and neurological examination showed no abnormality. Ophthalmologic examination showed binocular visual acuity of 20/33, and the binocular intraocular pressure was 15 mmHg. Visual field examination was normal. Pituitary hormone levels were within the normal reference range. Brain MRI showed a 29 mm×18 mm×20 mm irregular lesion in the sellar region extending into the right cavernous sinus area. The lesion was well-defined and appeared isointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images. After administration of contrast medium, the lesion showed remarkable homogeneous enhancement (**Figure 2**). Additionally, the internal carotid artery was wrapped by the tumor, and the optic chiasma was lifted. There was clear demarcation between the tumor and the hypophysis. A diagnosis of meningioma was suspected. Brain computed tomographic angiography (CTA) showed a nodular hyperdense lesion involving the right cavernous sinus, which had caused compres-

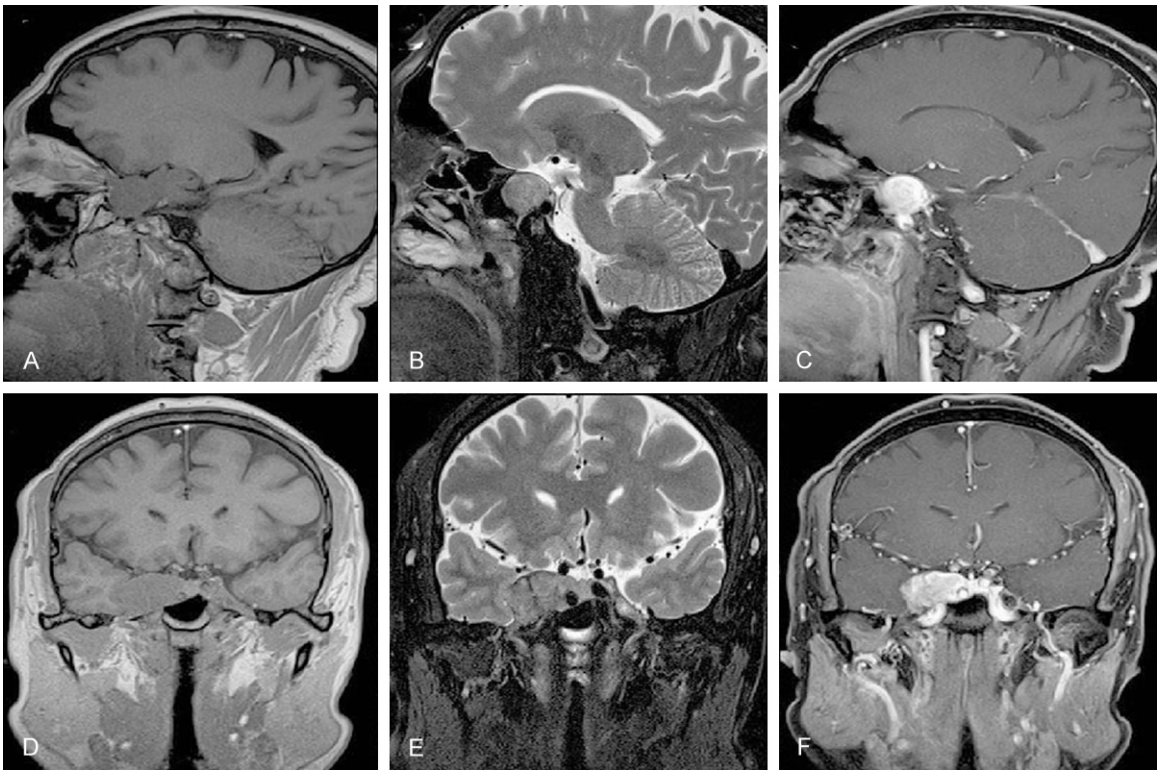

Figure 2. Magnetic resonance imaging of case 2. Sagittal (A) and coronal (D) T1-weighted images showing an isointense irregular lesion in the sellar region extending to the right cavernous sinus area. Sagittal (B) and coronal (E) T2-weighted images showing a heterogeneously hyperintense lesion. Sagittal (C) and coronal (F) T1-weighted contrast-enhanced images showing remarkable homogeneous enhancement of the lesion.

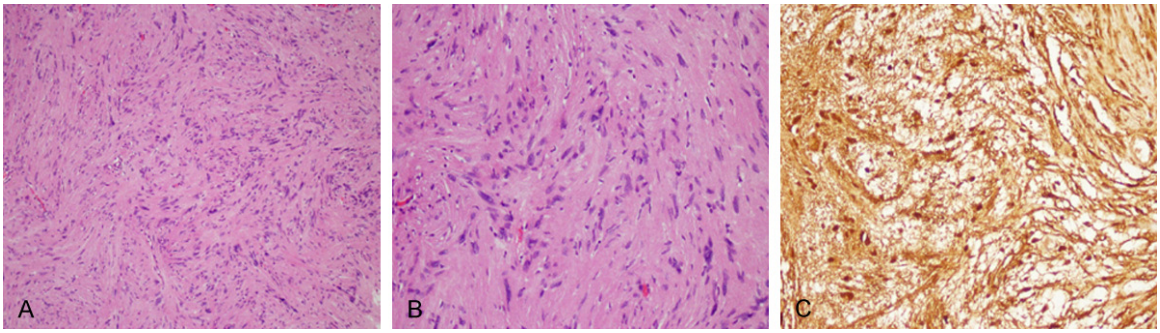

Figure 3. Pathological images of sellar schwannoma. Hematoxylin-eosin stained tumor sections (A, ×100; B, ×200) showing spindle-shaped Schwann cells arranged in interlacing fascicles; eosinophilic cytoplasm with poor demarcation, nuclear palisading, and thick-walled blood vessels are visible. Immumohistochemically stained tumor section showing cells positive for S-100 protein (C, ×200).

sional shift of the right siphon carotid artery. No significant feeding artery was identified. A surgical resection was performed via the right frontotemporal approach. Intraoperatively, a gray-white tumor was found located in the right intrasellar and parasellar region. No nerve of origin was identified. The entire tumor was removed in a piecemeal fashion. Histopathological examination confirmed the diagnosis of schwannoma (**Figure 3**). The patient performed well during the follow-up period.

Discussion

On a review of published literature, we identified a total of 30 reported cases of sellar sch-

Table 1. Clinical and radiological profiles of patients with schwannoma in sellar region

Author	Year	Gender	Age	Symptoms	Endocrine function	Radiological features	Treatment	Resection extent	Outcome
Chadduck et al. [9]	1973	F	53	Headache, status epilepticus	Normal	N.A.	Transcranial tumorectomy	N.A.	N.A.
Goebel et al. [10]	1979	F	25	Epilepsy	Mild hypogonadism	N.A.	Transcranial tumorectomy	GTR	Normal
Perone et al. [11]	1984	M	38	Headache	Hypopituitarism	N.A.	Trans-sphenoidal tumorectomy	GTR	Normal
Ishige et al. [12]	1985	F	64	Ptosis, diplopia	Normal	CT: enhancement	Trans-sphenoidal tumorectomy	GTR	Normal
Kasantikul et al. [13]	1987	F	48	Headache, visual field defect	Normal	N.A.	Transcranial tumorectomy	GTR	Subarachnoid hemorrhage
Wilberger et al. [7]	1989	F	62	Visual loss, hydrocephalus	Hypopituitarism	N.A.	Trans-sphenoidal tumorectomy + transcranial tumorectomy	Incomplete resection	Hypopituitarism, visual improvement
Guenot et al. [14]	1994	M	67	Headache, visual loss, oculomotor nerve paralysis	Hypopituitarism	MRI-T1: hypointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	STR	Visual improvement
Civit et al. [15]	1997	M	41	Bitemporal hemianopia	Normal	N.A.	Trans-sphenoidal tumorectomy + transcranial tumorectomy (3 months later)	GTR	Hypopituitarism, visual improvement, diabetes insipidus
Kim et al. [16]	2002	M	39	Nasosinusitis, bitemporal hemianopia	Hypopituitarism	N.A.	Trans-sphenoidal tumorectomy	STR	Normal
Whee et al. [2]	2002	M	39	Visual loss, hypaphrodisia	Hypopituitarism	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	STR	Hypopituitarism, visual improvement, diabetes insipidus
Bhagat et al. [17]	2002	M	68	Visual loss	Hypopituitarism	CT: extending to suprasellar region	Trans-sphenoidal tumorectomy	GTR	Hypopituitarism
		M	51	Hypaphrodisia, fatigue, somnolence	Hypopituitarism	Gd-DTPA: heterogeneously enhanced	Trans-sphenoidal tumorectomy	STR	Hypopituitarism
Maartens et al. [3]	2003	F	33	Headache, hypomenorrhea, visual loss	Normal	Gd-DTPA: heterogeneously enhanced	Trans-sphenoidal tumorectomy	GTR	Visual improvement
		F	56	Headache, bitemporal hemianopia, hydrocephalus	Normal	N.A.	Transcranial tumorectomy	STR	Hypopituitarism
Esposito et al. [18]	2004	M	73	Bitemporal hemianopia	Hypopituitarism	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	STR	Hypopituitarism, visual improvement
Perez et al. [19]	2004	F	71	Bitemporal hemianopia	Normal	MRI-T1: hypointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	PR	Visual improvement
Honegger et al. [20]	2005	F	79	Syncope	Normal	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	GTR	Visual improvement, diabetes insipidus
Moreland et al. [21]	2006	M	41	Headache, delirious, fatigue	Normal	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	STR	Normal
Rodriguez et al. [22]	2007	M	41	Facial pain, bitemporal hemianopia	Normal	Gd-DTPA: heterogeneously enhanced	Transcranial tumorectomy	PR	N.A.
Park et al. [4]	2009	F	49	Headache, vomiting, visual loss	Normal	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy + transcranial tumorectomy (5 months later)	GTR	Normal
Mohammed et al. [23]	2010	F	19	Visual field defect	Normal	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	GTR	Normal

		F	45	Headache, facial pain	Normal	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	GTR	Hypopituitarism
Koutourousiou et al. [24]	2010	M	38	Acromegalia	Elevated growth hormone	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	GTR	Normal
Boj-carcellar et al. [25]	2012	F	56	Headache	Normal	Gd-DTPA: heterogeneously enhanced	Trans-sphenoidal tumorectomy + transcranial tumorectomy	GTR	Hypopituitarism
Cugati et al. [26]	2012	M	42	Headache, blurred vision	Normal	MRI-T1: isointense MRI-T2: hyperintense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	GTR	Hypopituitarism
Senapati et al. [27]	2014	F	24	Ptosis, diplopia	Normal	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Transcranial tumorectomy	GTR	Normal
Sharifi et al. [6]	2015	F	45	Headache, facial pain, visual loss, bitemporal hemianopia	Normal	MRI-T1: hypointense MRI-T2: hyperintense Gd-DTPA: heterogeneously enhanced	Trans-sphenoidal tumorectomy	GTR	Normal
Kong et al. [1]	2015	F	65	Headache, fatigue, depression, visual loss	Hypopituitarism	MRI-T1: isointense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	PR	Normal
Zhang et al. [28]	2016	M	50	Headache, blurred vision	Hypopituitarism	MRI-T1: hyperintense Gd-DTPA: remarkably enhanced	Trans-sphenoidal tumorectomy	GTR	Hypothyroidism
Liu et al. [29]	2016	M	50	Visual loss	Normal	MRI-T1: isointense Gd-DTPA: heterogeneously enhanced	Trans-sphenoidal tumorectomy	GTR	Hypopituitarism
Present case 1	2017	F	51	Asymptomatic	Normal	MRI-T1: isointense MRI-T2: hyperintense Gd-DTPA: homogeneously enhanced	Trans-sphenoidal tumorectomy	STR	Normal
Present case 2	2017	M	50	Headache	Normal	MRI-T1: isointense MRI-T2: hyperintense Gd-DTPA: homogeneously enhanced	Transcranial tumorectomy	GTR	Normal

F, female; *M*, male; *N.A.*, not available; *GTR*, gross total resection; *STR*, subtotal resection; *PR*, partial resection.

wannoma. The clinical and radiological details of these cases are summarized in **Table 1**.

The 32 patients (including the current two cases) diagnosed with sellar schwannoma included 15 males (46.9%) and 17 females (53.1%). The male-to-female ratio was 1:1.1. The age at diagnosis ranged from 4-79 years [mean: 49.2 (±14.5) years].

The clinical manifestations included visual disturbance (n=27, 84.4%), headache (n=15, 46.9%), oculomotor nerve paralysis (n=3, 9.4%), dysendocriniasis (n=4, 12.5%), and epilepsy (n=2, 6.3%). On MRI, the sellar schwannomas appeared hypointense (3/18), isointense (14/18), or hyperintense (1/18) on T1-weighted images, and hyperintense (4/4) on T2-weighted images. All lesions (22/22) showed remarkable contrast-enhancement. All the 32 patients underwent surgical treatment, 7 of which received transcranial tumorectomy and 28 received trans-sphenoidal tumorectomy and 4 patients underwent both trans-sphenoidal tumorectomy and transcranial tumorectomy. Gross total resection was achieved in 20 out of the 32 (62.5%) patients. Postoperatively, endocrine complications were noted in 13 out of the 32 (40.6%) patients.

Intracranial schwannomas most frequently originate from sensory nerves such as the sensory branch of trigeminal nerve and the vestibular nerve and those occurring in the sellar region are exceedingly rare. The definitive pathogenesis of primary intrasellar schwannomas is not clear, especially the origin of sellar schwannomas remains obscure since none of the nerves traverse the sella turcica. Several theories pertaining to the pathogenesis and development of sellar schwannomas have been proposed. Maartens et al. proposed that sellar schwannomas may originate from the lateral sellar nerve plexus that innervates visceral motor and sensory nerves [3]. A second theory hypothesizes that sellar schwannomas may originate from hyperplastic Schwann cells of perivascular nerves or small dural sensory branches of the trigeminal nerve [4]. Another hypothesis links sellar schwannoma with abnormal cell proliferation or chronic disease of the Schwann cells [4]. Others have attributed these neoplasms to ectopic Schwann cells or to transformation of pial cells and pluripotential mesenchymal cells into Schwann cells [5]. In the current cases, no nerve of origin was identified. Further studies are warranted to elucidate the pathogenesis of sellar schwannoma.

The clinical manifestations of sellar schwannomas tend to be non-specific and similar to those of pituitary adenomas, which predominantly depend on the location and size of the tumor. The most common symptoms are visual disturbances caused by compression of optic nerve, including visual loss and bitemporal hemianopia. Extension of the tumor into the cavernous sinus region typically leads to oculomotor nerve involvement, which manifests as diplopia or ptosis. Compression of the pituitary stalk may lead to hypopituitarism. The space occupying effect of tumors can lead to headache, and tumors of large size can cause intracranial hypertension and even cognitive dysfunction [4].

Preoperative radiological diagnosis of sellar schwannomas is typically challenging. On computed tomography (CT), sellar schwannomas appear isodense with significant enhancement. On MRI, these entities usually appear hypo- to isointense on T1-weighted sequences and hyperintense on T2-weighted sequences. The lesions typically show remarkable homogenous contrast-enhancement. Calcification is uncommon, and has only been reported in 1 patient [6]. The differential diagnosis should include pituitary adenoma and sellar meningioma.

Definitive diagnosis is based on histopathological examination. Microscopically, schwannomas exhibit spindle-shaped Schwann cells arranged in interlacing fascicles with mild nuclear pleomorphism and no significant mitotic activity. On immumohistochemical staining, schwannomas stain positive for S-100 protein and epithelial membrane antigen (EMA), but stain negative for adenohypophysis markers. In the current cases, the histopathological findings were consistent with the above features.

Due to the rarity of sellar schwannomas, optimal treatment for these entities is not yet outlined. Considering the rich blood supply and hard nature of tumor tissues, Wilberger proposed that the transcranial approach may have an advantage over the trans-sphenoidal approach [7]. With advances in trans-sphenoidal microneurosurgery, the safety and total resection rate of intrasellar tumors has been

greatly improved. For tumors extending to suprasellar and/or parasellar regions, the transcranial approach may be more appropriate. Additionally, transcranial tumorectomy is also indicated for remnant or recurrent tumors. In the published literature, four patients underwent second-stage transcranial tumorectomy after failure of complete resection with transsphenoidal surgery. The role of adjuvant radiotherapy is not clear. Radiation is generally considered ineffective for schwannomas; however, Krayenbuhl reported a patient with intrasellar malignant peripheral nerve sheath tumor in whom fractionated conventional radiation of the sellar region helped achieve local disease control for 9 years [8].

Conclusion

Schwannomas arising in the sellar region are extremely rare entities. Preoperative differential diagnosis is typically challenging, and definitive diagnosis depends on histopathological and immunohistochemical evidence. Surgical resection is the treatment of first-choice. Surgical resection is the treatment of first-choice. Craniotomy is the most effective method for the treatment. Early detection and treatment can significantly improve clinical symptoms.

Disclosure of conflict of interest

None.

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