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Review Article

Ectopic schwannoma of the sellar region in a 1-year-old child: A case report and review of literature

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ABSTRACT

Background: Schwannomas are cranial and spinal nerves' sheath tumors accounting for up to 8% of all intracranial neoplasms. Although typical intracranial schwannomas originate from Schwann cells surrounding cranial nerves, ectopic schwannomas are not associated with a known cranial nerve or have an unknown origin. The location of schwannomas may impose clinical challenges. Sellar region schwannomas are rare whether it is ectopic or not. Herein, we report a pediatric case of a 1-year-old female with ectopic, intra-supra sellar with a literature review. We report the first case of juvenile ectopic schwannoma in the sellar region.

Methods: A PubMed Medline database search was performed by the following combined formula of medical subject headings (MESH) terms and keywords: ((sella turcica [MeSH Terms]) OR (sella*[Title/Abstract]) OR (ectopic [Title/Abstract]) AND ((neurilemmoma [MeSH Terms]) OR (schwannoma [Title/Abstract]) OR (neuroma [Title/Abstract]) OR (neurinoma [Title/Abstract])).

Results: Total results of 206 articles were obtained. In exclusion of intraparenchymal and intraventricular schwannomas, only 34 articles remained. Thirty-nine cases were included in 34 articles. According to the reported cases, intrasellar schwannomas are more common in elderly individuals in an average of 49.5 years (range: 19–79 years). They have a good prognosis and affect males to females equally (20:19).

Conclusion: Ectopic schwannoma sited in the sellar region is rare. It is the first case to be reported in the pediatric age group with a literature review. This lesion should be highlighted and included in the differential diagnosis of sellar mass.

Keywords: Ectopic schwannoma, Neuroma, Pediatric, Sellar tumors

INTRODUCTION

Schwannomas are common cranial and spinal nerves' sheath tumors representing up to 8% of all intracranial neoplasms.[28] Typical intracranial schwannomas are originating from Schwann cells surrounding cranial nerves' axons and are considered Grade I benign tumors. The vast superiority of cranial schwannomas is associated with the vestibulocochlear nerve (CN VIII) in cerebellopontine angle as acoustic neuroma.[34] Intracranial schwannomas that are not associated with a known cranial nerve or have an unknown origin are called ectopic schwannomas. There are many hypothesized theories about their origin. When located in the sellar region, schwannomas often are misdiagnosed with pituitary adenomas (PAs) and are not considered nonadenomatous

sellar lesion, which carries a high intraoperative risk due to their high vascularity.[31] Whether ectopic or nonectopic, sellar schwannomas are a rare entity. To the best of our knowledge, our case is the first juvenile ectopic schwannoma in the sellar region to be reported in the literature.

Herein, we report a pediatric case of a 1-year-old female with ectopic, intra-supra sellar, relatively large schwannoma associated with severe visual impairments, weakness, and endocrine disturbances. She was treated with a subtotal tumorectomy and followed up for 1 year. Good prognosis and noticed improvement of visual and endocrine states proved by postoperative examination. In addition, we conducted a review of all sellar schwannomas reported in the literature.

CASE SCENARIO

A 1-year-old female was admitted to our hospital with complaints of severe visual impairments in both eyes, strabismus, weakness, and vomiting. She also had endocrinal disturbances, including hyperthyroidism (Free thyroxine: 26.08 pmol/L and free triiodothyronine: 8.08 pmol/L). Preoperative Magnetic Resonance Imaging (MRI) [Figure 1] revealed hyperintense mass (40*35*30) mm located in the sellar region and extending suprasellar with third ventricle, hypophysis, and optic chiasm compression and minimal lateral and third ventricles dilation.

The patient underwent transcranial tumorectomy with a right-sided pterional approach. Subtotal removal of a rubbery high vascularized mass was achieved, and no postoperative complications were exacerbated. She had gradual improvement of vision and endocrine function postoperatively and regained full vision within 1 year of follow-up [Figure 2]. Histopathology manifested a schwannoma with positive staining for S-100 protein and negative for glial fibrillary acidic protein and human epithelial membrane antigen [Figure 3].

METHODS

We conducted a PubMed Medline database search by the following combined formula of medical subject headings (MESH) terms and keywords: ((sella turcica [MeSH Terms]) OR (sella*[Title/Abstract]) OR (ectopic [Title/Abstract]) AND ((neurilemmoma [MeSH Terms]) OR (schwannoma [Title/Abstract]) OR (neuroma [Title/Abstract]) OR (neurinoma [Title/Abstract])).

RESULTS

A total of 206 results were obtained, and by the exclusion of intraparenchymal and intraventricular schwannomas, only 34 articles remained. Thirty-nine cases were included in 34 articles. According to the reported cases [Table 1], intrasellar

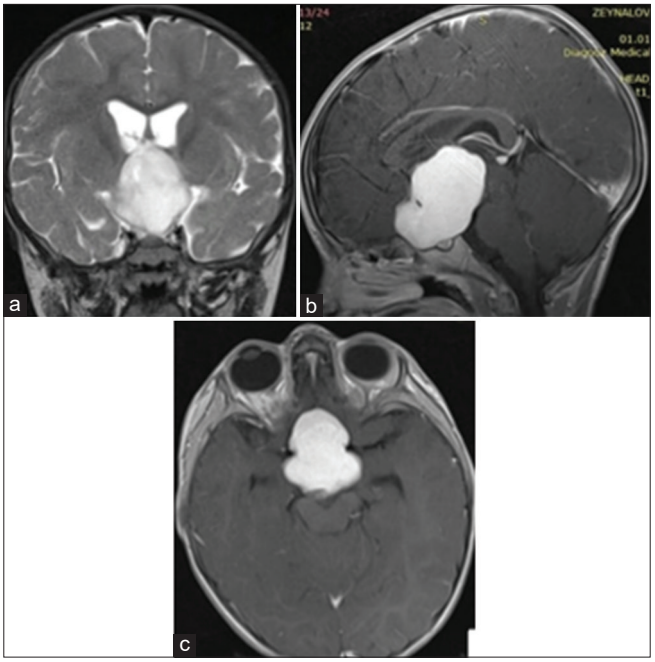

Figure 1: Preoperative magnetic resonance image: (a) T2-weighted image (coronal section) demonstrates hyperintense mass localized in the sella and extending to the suprasellar area, compressing the third ventricle. (b) T1-weighted image (B: sagittal), a contrast-enhanced image of the brain showing homogenous enhancement of the lesion. (c) T1-weighted image (axial section), contrast-enhanced image showing homogenous enhancement of the mass.

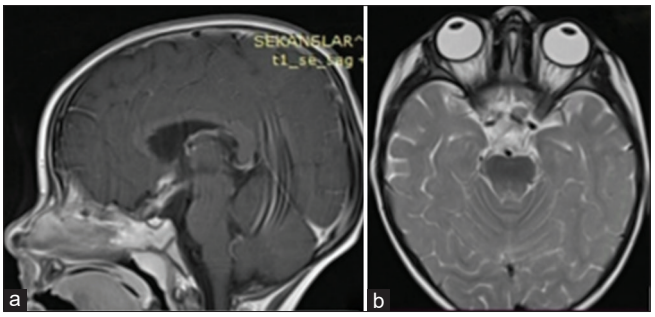

Figure 2: One-year follow-up magnetic resonance image: (a) T1-weighted image (sagittal section), affirm successful tumorectomy and decompression of optic chiasm with normal ventricular volume. (b) T2-weighted image (axial section), confirm successful tumorectomy and decompression of optic chiasm with no dilatation of the ventricles.

schwannomas pick young to elder age groups and are more common in elderly individuals in an average of 49.5 years (range: 19–79 years).[1,3-9,11-16,19,21-25,27,29,30,32,33,35] They have a good prognosis and affect males to females equally (20:19).

DISCUSSION

The significant preponderance of sellar lesions is for PAs. Other nonadenomatous sellar lesions include neoplastic, cystic,

Table 1: Literature review of Sellar schwannomas cases.

Authors, Year of Publication	Gender, Age (years)	Symptoms and Signs	Endocrine Function	Surgical Approach	Outcome
Chadduck[4] 1973	F* 53	HA*, status epilepticus	Normal	TCA*	Expired after surgery
Goebel *et al.*[8] 1979	F 25	Epilepsy	Mild hypogonadism	TCA	Postoperative seizure
Perone *et al.*[27] 1984	M 38	HA	Hypopituitarism	TSA*	Transient, minimally decreased left corneal reflex
Ishige *et al.*[12] 1985	F 64	Ptosis, diplopia	Normal	TSA	Facial pain improvement
Kasantikul *et al.*[13] 1987	F 48	HA, visual field defect	Normal	TCA	Transient, minimally decreased right corneal reflex
Wilberger *et al.*[33] 1989	F 62	Visual loss, hydrocephalus	Hypopituitarism	TSA+TCA	Hypopituitarism, Continued visual loss
Guenot *et al.*[9] 1994	M 67	HA, visual loss, oculomotor nerve paralysis	Hypopituitarism	TSA	Visual improvement
Civit *et al.*[5] 1997	M 41	Bitemporal hemianopia	Normal	TSA+TCA (3 months later)	Hypopituitarism, visual improvement, diabetes insipidus
Kim and Suh[14] 2002	M 39	Nasosinusitis, bitemporal hemianopia	Hypopituitarism	TSA	Visual improvement
Whee *et al.*[32] 2002	M 39	Visual loss, hypaphrodisia	Hypopituitarism	TSA	Hypopituitarism, visual improvement, transient diabetes insipidus
Bhagat *et al.*[2] 2002	M 68	Visual loss	Hypopituitarism	TSA	Visual improvement
	M 51	Hypaphrodisia, fatigue, somnolence	Hypopituitarism	TSA	Hypopituitarism
Maartens *et al.*[21] 2003	F 33	HA, hypomenorrhea, visual loss	Normal	TSA	Visual improvement
	F 56	HA, bitemporal hemianopia, hydrocephalus	Normal	TCA	Continued hydrocephalus, ventriculoperitoneal shunt insertion
Esposito *et al.*[7] 2004	M 73	Bitemporal hemianopia	Hypopituitarism	TSA	Hypopituitarism, visual improvement
Perez *et al.*[26] 2004	F 71	Bitemporal hemianopia	Normal	TSA	Visual improvement after surgery Profuse bleeding during surgery (5 units pRBC transfusion)
Honegger *et al.*[11]2005	F 79	Syncope	Normal	TSA	Visual improvement, diabetes insipidus
Yoon *et al.*[34] 2005	M 34	HA	Hypothyroidism	6 months thyroid hormone therapy, TSA biopsy, and resection	Normalized thyroid function
	M 62	HA & visual disturbance & polyuria with polydipsia	Not defined	TSA	Visual symptoms and diabetes insipidus both improved
Moreland[23] 2006	M 41	HA, delirious, fatigue	Normal	TSA	Normal
Rodriguez *et al.*[29] 2007	M 41	Facial pain, bitemporal hemianopia	Normal	TCA	Hyperglycemia, low thyrotropin levels, and hyponatremia
Park *et al.*[25] 2009	F 49	HA, vomiting, visual loss	Normal	TSA+TCA (5 months later)	Normal

(Contd...)

Table 1: *(Continued).*

Authors, Year of Publication	Gender, Age (years)	Symptoms and Signs	Endocrine Function	Surgical Approach	Outcome
Mohammed *et al.*[22] 2010	F 19	Visual field defect	Normal	TSA	Vasospasm, bifrontal infarction
	F 45	HA and facial pain	Hypopituitarism	TSA	Vasospasm, bifrontal infarction Mild hypopituitarism
Koutourousiou *et al.*[16] 2010	M 38	Acromegalia	Elevated growth hormone	TSA	Normalized pituitary function
Boj-carcellar *et al.*[3] 2012	F 56	HA	Normal	TSA+TCA	Hypopituitarism
Cugati *et al.*[6] 2012	M 42	HA, blurred vision	Normal	TSA	Visual improvement
Luo *et al.*[19] 2013	M 38	Acromegaly, headache and decreased libido	Increased growth hormone	TSA	Remission of acromegaly Resolved headache
Puduru[28] 2013	F 67	Bitemporal hemianopia	Hypopituitarism	TSA	Hypocortisolism
Senapati *et al.*[30] 2014	F 24	Ptosis, diplopia	Normal	TCA	Normal
Sharifi *et al.*[31] 2015	F 45	HA, facial pain, visual loss, bitemporal hemianopia	Normal	TSA	Visual improvement, facial pain improvement
Zhang *et al.*[36] 2016	M 50	HA, blurred vision	Hypopituitarism	TSA	Hypothyroidism
Kong *et al.*[15] 2016	F 65	HA, fatigue, depression, visual loss	Hypopituitarism	TSA	No headache, relieved visual field, improved pituitary function
Liu *et al.*[17] 2016	M 50	Visual loss	Normal	TSA	Hypopituitarism
Bae *et al.*[1] 2018	F 57	Incidental finding on screening MRI		TCA	Profuse bleeding during surgery, permanent clipping and coil embolization sacrifice of carotid artery, postoperative cerebral infarction in the left basal ganglia
Oishi *et al.*[24] 2017	M 64	Gait disturbance, left lateral hemianopia	Hypopituitarism	TCA	Improvement of gait and vision
Zhang *et al.*[35] 2018	F 51	Asymptomatic	Normal	TSA	Normal
	M 50	HA	Normal	TCA	Headache improvement
Lyu *et al.*[20] 2021	M 38	HA, bilateral supratemporal quadrantopsia.	Hypopituitarism	TSA	Normalized pituitary function
Present case 2022	F 1	Severe visual impairments in both eyes, strabismus, weakness	Hyperthyroidism	TCA	Visual improvement

HA: Headache, F: Female, M: Male, TCA: Tanscranial tumorectomy, TSA: Tans-sphenoidal tumorectomy

and inflammatory lesions.[1] Even though the radiological characteristics of each are well known, they happen to be indistinguishable. schwannomas, also called neuromas or neurilemmoma, are common intracranial nerve sheath tumors related to a genetic syndrome called neurofibromatosis-2.[10] However, they are scarcely located in the sellar region and are frequently misdiagnosed with PAs as they are not considered in their differential diagnosis due to their rarity.

The clinical presentation of sellar schwannomas is represented by the triad: headache, visual disturbances, and endocrinal dysfunction; most commonly hypopituitarism which is believed to be a result of pituitary stalk and portal vessels compression.[26] Intrasellar schwannomas' radiological findings on computed tomography include isodense mass with homogenous enhancement and isointense (T1WI), slightly hyperintense (T2WI) on MRI with homogenous, or heterogeneous enhancement.[17] The final declaration of the diagnosis is based on histopathology, and immunohistochemical examination as their reaction is intense with S-100 and vimentin.[36] Intrasellar schwannomas are mostly extending suprasellar and may coexist with other lesions or mimic other intra-axial tumors.

Although schwannomas are benign tumors of children and adults, sellar schwannomas have not been reported in

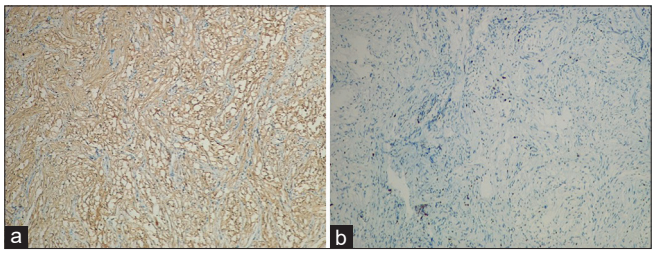

Figure 3: (a) A histopathological image shows spindle cells with immunostaining that is positive staining for S-100 protein and negative for glial fibrillary acidic protein and epithelial membrane antigen. It is consistent with Schwannoma, the WHO Grade 1. (b) A histopathological image of Schwannoma with nuclear expression of Ki67 proliferation index was observed in approximately 2–3% of cells.

pediatrics yet. Our case is the first juvenile intra-suprasellar schwannoma, and the clinical presentation is the same with adults, in addition to weakness and vomiting.

According to their origin, schwannomas are peripheral nervous system tumors arising from Schwann cells ensheathing peripheral axons of all cranial nerves except the optic nerve (CN II). Their most common sites are associated with the vestibulocochlear nerve (CN VIII) in acoustic neuroma, second with the trigeminal nerve (CN V).[34] They are described as ectopic if they had no relation to any of the known cranial nerves or if they arose of unknown origin.[18] Ectopic intracranial schwannomas can be intraparenchymal, intraventricular, and very rarely intrasellar. As Schwann cells are not innate central nervous system components, four hypothesized histopathological theories about schwannomas' origin have been suggested:[20] (1) Schwann cells ensheathing the tiny nerve twigs which innervate the dura, (2) ectopic Schwann cells, (3) perivascular Schwann cells (e.g., perivascular nerve plexus are encasing the internal carotid artery wall[1]), and (4) cranial nerves of the cavernous sinus (e.g., lateral sellar nerve plexus). In our case, the most reasonable presumptive origin of schwannoma is ectopic Schwann cells in the sellar area.

In our literature review, Chadduck, in 1973, first reported the case of a 53-year-old female who presented with a headache; then, she developed an attack of status epilepticus. Her management included transcranial tumorectomy (TCA); nevertheless, she expired postoperatively. Thirty-eight cases then were followed in the reporting of this pathology. None of the reported cases were within the pediatric age group, and the youngest person was 19-year-old, and the case was reported by Mohammed *et al*. 2010. The current case is the first to be reported as an infant and within the pediatric age group.

In summary, a 53-year-old female presented with visional impairment and vomiting. Her imaging showed a mass in the sellar region. The surgery was considered, and a TCA was performed. In the follow-up period, the patient fully recovered in the vision and endocrine function in 1 year. This case is unique and solo as it is the first juvenile ectopic schwannoma.

CONCLUSION

Ectopic schwannoma located in the sellar region is rare. It is the first case to be reported in the pediatric age group with a literature review. This pathology should be included in the differential diagnosis of the mass in the sellar region, particularly when the radiological features of the mass are suspicious.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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