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Brainstem schwannoma: A case report and review of clinical and imaging features

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► ***Case report***

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ABSTRACT

Background: Intracerebral schwannoma is an extremely rare disease, accounting for fewer than 1% of intracranial schwannomas. The most common site for this type of schwannoma is the cerebral hemisphere, especially the frontal and temporal lobes; brainstem schwannoma is infrequent. ***Case Description:*** Here, we report a 51-year-old man with a monthlong history of blurred vision and weakness in his left lower limb. Magnetic resonance imaging showed a heterogeneous cystic tumor with a solid nodule arising from the brainstem. The patient underwent a craniotomy with complete resection of the tumor, which was confirmed to be a brainstem schwannoma by histopathological examination. We also performed a literature review of the 19 reported cases of brainstem schwannoma. ***Conclusions:*** Brainstem schwannomas predominated in children and young adults (60% of cases occurred in patients ≤ 30 years of age), and were more common in females (65%). Most of these schwannomas exhibited heterogeneous intensity, containing cystic (78%) and solid-enhanced components. The vast majority of reported cases (94.9%) followed a benign course, with an improved prognosis following tumor resection.

Keywords: *Brainstem schwannoma, clinical and imaging features.*

INTRODUCTION

Intracranial schwannoma accounts for 5–8% of intracranial tumors, whereas intracerebral schwannoma, a rare disease, accounts for <1% of intracranial schwannoma cases [1]. Since the first case of brainstem schwannoma was described in 1980 [2], only 19 cases [2-16] have been reported to date (table 1). This study reports a new case, along with a review of the clinicopathological and radiological characteristics of brainstem schwannoma, which may prove helpful in improving the capacity to image and clinically diagnose this disease [16].

Case report

A 51-year-old man developed blurred vision and weakness in his left lower limb lasting 1 month. The symptoms continued to worsen without nausea, vomiting, or headache. Physical examinations were negative for the left-hand alternating movement, heel-knee-tibia, Romberg, and finger-nose tests. The patient underwent preoperative magnetic resonance imaging (MRI) examination. , which showed a heterogeneous cystic lesion with a solid nodule involving the midbrain and pons. The cystic component was hypointense on T1-weighted image (T1WI) and hyperintense on T2-weighted image (T2WI); the solid nodule was isointense on T1WI, T2WI, and diffusion-weighted imaging (DWI). The tumor was heterogeneously enhanced after contrast agent administration (figure 1).

The patient underwent a craniotomy for tumor resection. After the arachnoid sheath had

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been dissected, a firm and gray-yellow tumor with rich vascularity originating from the brainstem was observed, measuring 3 cm × 4 cm × 3.5 cm. Microscopic examination revealed a spindle cell neoplasm characterized by Antoni A and Antoni B areas, with Verocay body's characteristic of schwannomas. Immunohistochemical analyses of protein S-100 and SRY-related HMG-box 10 (SOX10) reactivities were strongly positive for tumor cells, but negative for glial fibrillary acidic protein (GFAP). In addition, the tumor cells were immunopositive for Ki67 (1 %+) and cluster of differentiation 34 (CD34) (vascular+) (figure 2). The surgical and histopathological findings confirmed the diagnosis of schwannoma arising from the brainstem.

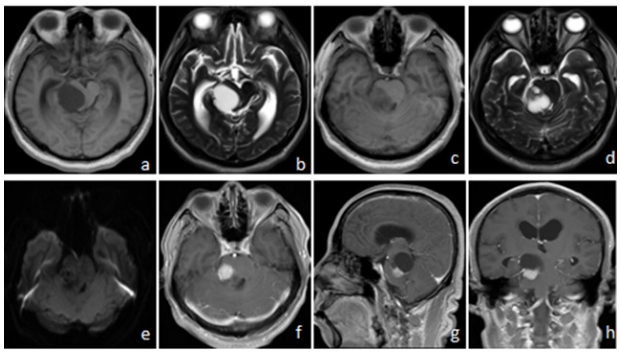

Figure 1. Precontrast MRI showing that the cystic component was hypointense on T1WI **(a)**, hyperintense on T2WI **(b)**, and the solid nodule was isointense on T1WI **(c)**, T2WI **(d)**, and DWI **(e)**. Contrast MRI showing that the mass was heterogeneously enhanced **(f, g)**.

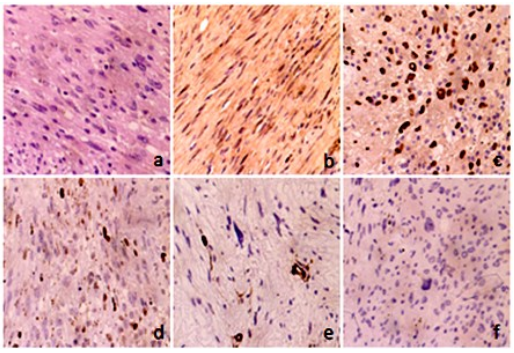

Figure 2. Microscopic images showing **(a)** hematoxylin and eosin areas of nuclear palisading (200×). Photomicrographs of immunohistochemical analysis showing **(b)** strong positivity to S100 staining (200×), **(c)** intense positive staining to SOX10 (200×), **(d, e)** weak positivity to Ki67 (200×) and CD34 (200×), and (f) negativity to glial fibrillary acidic protein (GFAP) (200×).

DISCUSSION

Intracerebral schwannoma, a rare disease, accounts for <1% of intracranial schwannoma cases. The most common sites are the frontal and temporal lobes of the cerebral hemispheres [9]; they have also been found in the cerebellar hemispheres and vermis [17] in relation to the tentorium cerebelli and the lateral and fourth ventricles. Prakash *et al.* [2] first reported a case of a 14-year-old female with a schwannoma in the brainstem, which was successfully surgically resected in 1980. To date, only 19 cases of schwannoma originating in the brainstem have been reported. Based on a review of the literature, we summarized the clinicopathological and imaging features of brainstem schwannomas.

Intracerebral schwannomas are quite rare, as Schwann cells are not normally found in the brain or spinal cord parenchyma. The clinical features of all known brainstem schwannomas are presented in table 1. According to our literature review, the age of affected patients ranged from 12 to 68 years (average age: 31.7 years), and this type of tumor was more likely to occur in children and young adults (60% of cases occurred in patients ≤ 30 years of age). The male-to-female ratio was 1:1.86, and females accounted for 65% of the cases, indicating that women may be more frequently affected than men. The symptom duration varied from 6 weeks to 3 years, with a median duration of 7 months. Brainstem schwannomas lack specific clinical manifestations, as the majority of patients experienced blurred vision, headache, nausea, monoparesis, and gait disturbances.

The typical imaging characteristics of brainstem schwannomas were a well-circumscribed mass with cyst formation and a solid enhanced component [18]. Computed tomography (CT) indicated an iso-density or low -density contrast-enhancing mass with a cyst. The tumors were hypointense and hyperintense on T1WI and T2WI, respectively [17]. The solid

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component was strongly enhanced after contrast agent injection. In our case, the tumor was a heterogeneous cystic lesion with an enhanced solid nodule**.** With the exception of cases 1 and 9, which had restricted data, cystic degeneration was documented by preoperative CT/MRI images or intraoperative descriptions in 14 (78%) of the remaining 18 cases (table 2). Some researchers believe that the pathogenesis of cyst formation involves small cysts within solid tumors derived from sites of microhemorrhage, necrosis, or degeneration, which fuse to become a larger cystic component [18]. Compression of the brainstem and adjacent brain parenchyma by the tumor may result in mild edema**,** although the cause of edema around this benign neoplasm is uncertain. Moreover, calcification was rare. Gao *et al.* [16] reported a 12-year-old female whose CT images exhibited calcification shadows, but none of the other case studies contained explicit references to calcification.

Table 1. The 20 reported cases of brainstem schwannoma: clinical and surgical features.

NO.	References	Gender	Age	Site of tumor	Symptoms	Duration of symptons (months)	Treatment	Result
1	Prakash, 1980	F	14	Pons	Facial asymmetry, squint, tinnitus, vertigo, 6th & 7th cranial nerve palsies, ataxia	36	decompression	good
2	Aryanpur and Long, 1988	F	50	Medulla oblongata	Nausea, emesis, left -side limb numbness, diplopia, slight slurring of speech, unsteadiness of gait	1.5	excision	excellent
3	Ladouceur, 1989	F	46	brainstem	Blurred vision, dysarthria, dysphagia, Left-side weakness and gait unsteadiness evolving	6	excision	excellent
4	Sharma and Newton, 1993	M	18	Medulla oblongata	NA	24	NA	NA
5	Tanabe, 1996	F	68	Midbrain and pons	Right hemiparesis, sensory disturbance, and diplopia	7	excision	excellent
6	Sharma, 1996	M	14	Brainstem	Unsteadiness of gait, decreased vision both eyes, abnormal behaviour	12	excision	Be lost to follow up
7	Sharma, 1996	M	14	pons	Squint and facial asymmetry, Unsteadiness, 6th and 7th nerve paresis, left side ataxia	3	excision	good
8	Lee, 1999	F	29	Brainstem, cerebellum, spinal cord	Facial hypoesthesia, insufficiency of CN VII, monoparesis	36	NA	NA
9	Wang H, 2001	F	16	brainstem	Fever, cough, bucking, right-side weakness	24	Partial excision	Be lost to follow up
10	Lin J, 2003	M	48	Medulla oblongata	Right hemiparesis, ataxia and dysphagia	24	excision	excellent
11	Muzzafar, 2010	M	68	Midbrain, Midbrain, pons, and medulla	Gait imbalance, coughing, and hiccups, nausea, vomiting, intermittent diplopia, and weakness in right arm	2	excision	excellent
12	Srivastav, 2011	M	13	Midbrain and pons	Left hemiparesis, ataxia and slurring of speech, headache	4	excision	excellent
13	Ramos, 2013	F	17	Midbrain and pons	dizziness, unsteadiness, and headache, Left nystagmus, facial weakness, VI and VII nerves palsy, diplopia, and ataxia	3	excision	excellent
14	Konovalov, 2013	F	44	Midbrain	Left-sided numbness,	7	excision	excellent

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Continuation of Table 1. The 20 reported cases of brainstem schwannoma: clinical and surgical features.

15	Konovalov, 2013	F	22	Fourth ventricle brainstem	Intense headache, diplopia, Unsteadiness in walking, weakness of facial muscles on the left side, taste change, and left-sided hearing deterioration	3	excision	good
16	Konovalov, 2013	F	23	Medulla oblongata	Weakness and numbness of the right half of the body	2	excision	excellent
17	Sharma, 2016	F	26	Pons and medulla oblongata	Headache, progressive gait ataxia, right hemiparesis, left facial paresis, and slurring of speech	7	excision	excellent
18	Zhang Q, 2016	F	40	Medulla oblongata and cervical spinal cord	Cervical pain, weakness of the upper extremities and glove distribution numbness	12	excision	excellent
19	GAO Y, 2016	F	12	Medulla oblongata	Dizziness, headache, nausea, vomiting and require assistance with walking	12	excision	excellent
20	Present study	M	51	Brainstem	Blurred vision, left lower limb weakness	1	excision	excellent

M, male; F, female; NA, not available.

Table 2. Imaging features of the reported cases of brainstem schwannoma.

NO.	References	Radiological feature	Cystic
1	Prakash,1980	PEG and ventriculogram suggestive of a firm and abnormally vascular mass	No
2	Aryanpur, 1988	A hypointense on T1WI and hyperintense on T2WI mass with surrounding areas of iso-intensity	Yes
3	Ladouceur, 1989	A complex mostly cystic mass, the solid portion were enhanced obviously	Yes
4	Sharma,1993	NA	NA
5	Tanabe, 1996	A mixed signal in both T1WI and T2WI and heterogeneously enhanced tumor with cystic component	Yes
6	Sharma, 1996	Enhancing mass in right lower medulla	No
7	Sharma, 1996	PEG and ventriculogram suggestive of positive mass	No
8	Lee, 1999	NA	No
9	Wang H, 2001	Ventriculogram show a fourth ventricular mass	NA
10	Lin J, 2003	A partially cystic lesion surrounded by a ring of intense contrast enhancement	Yes
11	Muzzafar, 2010	A cystic septate lesion, and had enhancing septa	Yes
12	Srivastav, 2011	A lesion, hypointense on T1WI and homogenously hyperintense on T2WI, intense enhancement of the rim of the tumor with some solid part	Yes
13	Ramos, 2013	A heterogeneous cystic lesion, hypointense on T1WI, hyperintense on T2WI and had an enhancement of the rim of the tumor with an inferior mural nodule	Yes
14	Konovalov, 2013	A cystic neoplasm accumulating contrast	Yes
15	Konovalov, 2013	A contrast-accumulating cystic tumor	Yes
16	Konovalov, 2013	A bulk solid cystic tumor with the contrast agent being accumulated in the solid portion of the tumor	Yes
17	Sharma, 2016	An intra-axial, solid-cystic lesion, hypointense on T1WI, hyperintense on T2WI and showed enhancement	Yes
18	Zhang Q, 2016	A dumbbell-shaped mass, the nodular was enhanced obviously	Yes
19	GAO Y, 2016	An abnormal cystic signal and parenchyma parts presented nodular iso-intensity shadows above the cystic wall, CT showed calcified shadow	Yes
20	Present study	Heterogeneously enhanced tumor with cystic component	Yes

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Unfortunately, it is rather difficult to differentiate schwannomas from gliomas and hemangioblastomas using MRI, as gliomas are the most common tumors in the brainstem and all three lesions generally display similar MRI signals [9]. In fact, the neoplasm in our report was preoperatively diagnosed as a hemangioblastoma.

Typical schwannomas exhibit two types of tissue conformations: areas of spindle-shaped cells with elongated nuclei and eosinophilic cytoplasms (Antoni A), alternating with loose-textured areas of low cellular density associated with cyst formation (Antoni B) [17-20]. In our case study, upon immunohistochemical analysis, the tumor cells exhibited intense S-100 positivity, thereby confirming the diagnosis of schwannoma. Strongly expressed SOX10 may reveal the origin of the intracerebral schwannoma. Immunohistochemical staining of intracerebral schwannomas showed negative expression of epithelial membrane antigen and GFAP, distinguishing them from meningiomas and gliomas, and the proliferation index of Ki-67 did not increase (1% positive expression), indicating slow tumor cell proliferation.

In the vast majority of reported cases (93.7%), researchers apparently assumed that the intracerebral schwannomas were benign neoplasms, and no recurrence occurred following complete resection of the lesion. In one case, Zhang *et al*. [15] reported a 40-year-old woman who had tumor recurrence 20 months after the initial resection, although the prognosis was favorable following a second round of surgery. Apart from cases 1 and 8, which had restricted data, and cases 6 and 9, which lacked follow-up, the prognosis for the majority of the cases reviewed was classified as good. The postoperative course was uneventful for all of the patients and no new symptoms developed. Some patients, however, experienced sequelae after the operation including occasional transitory dizziness [4], mild dysesthesia of the right hand [6], and a trace of left fourth-nerve weakness [10].

CONCLUSION

We reported a 51-year-old male with brainstem schwannoma, a relatively rare condition, exhibiting distinctive imaging features that will help to improve preoperative diagnostic accuracy. In addition, the vast majority of reported cases (94.9%) followed a benign course, with an improved prognosis following tumor resection.

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Ethics approval and consent to participate

The present study was approved by the Ethics Committee of the First Clinical Medical College of Shanxi Medical University.

Conflicts of interest**:** *Declared none.*

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