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Asymptomatic Purely Intracranial Vagal Schwannoma: Clinical Case Report and Literature Review

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ABSTRACT

Vagus nerve schwannoma is an infrequently occurring schwannoma, in which a distinct subtype exists wherein the tumor is confined to the cerebellomedullary cistern without invading the jugular foramen. This unique tumor is called purely intracranial vagal schwannoma.

In this report, we present a case of purely intracranial vagal schwannoma in its asymptomatic early phase, incidentally discovered during surgery performed on a patient with hemifacial spasm. Because of the small size of the tumor, we definitively recognized that it originated from the second rootlet on the caudal side. The tumor was totally resected uneventfully and a favorable prognosis was achieved. Furthermore, we conducted a comprehensive literature review to summarize the classification, origin, and surgical complications associated with this rare tumor type.

Based on our literature review, we propose that: 1) the origin of tumor is related to the time of onset of symptoms, 2) nearly all purely intracranial vagal schwannomas can be entirely resected and favorable prognosis can be achieved, and 3) surgeons should be aware of potential cardiovascular complications during surgical procedures.

KEY WORDS: Vagal schwannoma, Intracranial, Jugular foramen, Asymptomatic

INTRODUCTION

Vagus nerve schwannoma is a rare tumor, mainly occurring in the jugular foramen and extracranial areas. However, some of these tumors are confined only to the cerebellomedullary cistern without any connection to the jugular foramen area; such tumors are referred to as purely intracranial vagal schwannoma. Based on previous findings in literature, only 10 cases of purely intracranial vagal schwannoma with confirmed pathological diagnosis have been reported; all of these exhibited evident compression symptoms (4-8,13,16-18). In this report, we present a case of purely intracranial vagal schwannoma that was discovered incidentally during surgery in a patient with hemifacial spasm who exhibited no other symptoms. Furthermore, we performed a literature review to enhance our understanding of diagnosis and treatment of this particular type of tumor.

CASE PRESENTATION

A 59-year-old woman had been experiencing paroxysmal, involuntary facial twitching on the left side of her face for the past ten years. Despite receiving drug therapy and acupuncture treatment, her symptoms had not improved. Upon admission, neurological examination revealed a left hemifacial spasm, while all other cranial nerve examinations were normal. An ECG revealed bradycardia, but the patient exhibited no cardiac symptoms. Furthermore, a preoperative CT scan did not indicate any significant abnormalities. However, a contrast-enhanced MRI identified a point of contact between the left facial nerve and a small artery. Diagnosis of primary hemifacial spasm was established, and retrosigmoid suboccipital craniotomy was performed. During the operation, we observed that the facial nerve was being compressed by the anterior inferior cerebellar artery at the root exit zone. Subsequently, we successfully separated the facial nerve from

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the artery using a nerve pad. Surprisingly, we also identified a grain-sized neoplasm attached to the vagus nerve located on the second rootlet from the caudal side (Figure 1). After obtaining consent from the patient's family, we excised the tumor-bearing rootlet and removed the tumor uneventfully. Postoperative pathology confirmed the diagnosis of benign schwannoma. Following the procedure, the hemifacial spasm disappeared immediately without any newly occurring neurological deficits. Three months after discharge, the patient recovered fully and did not report any discomfort during the follow-up examination.

LITERATURE REVIEW

Systematic literature searach of electronic databases, identified a total of 11 cases of vagus nerve schwannoma, including this case. Among these studies, one study was published full-text in Chinese and the others in English (Table I). The gender distribution was 4 males and 7 females, with a mean age of 56 years. Eight reports confirmed that the patients' diagnoses were not associated with neurofibromatosis, while in the remaining 3 reports this aspect was not mentioned. Except the patient in our case report, all the others exhibited tumor compression symptoms. The most common initial symptoms were related to lower cranial nerve dysfunction, and rare symptoms included changes in blood pressure. Within the cerebellomedullary cistern, the vagus nerve contains three distinct functional fiber types that are divided into several rootlets with specific distributions: pure sensory fibers are located in the rostral portion, motor/sensory fibers tend to be distributed towards the caudal section, and parasympathetic fibers are situated in the farthest caudal portion (1). Four cases demonstrated tumors in the rostral rootlets, one case showed a tumor in the caudal rootlet; however, the location of tumors was not mentioned in the other cases. All reports except one revealed that the tumors were totally resected after operation. Most intraoperative conditions were uneventful, although two patients experienced transient bradycardia and even asystole. Postoperative complications, including hoarseness, dysphagia, dysphonia, meningitis, and other symptoms were not uncommon.

DISCUSSION

Schwannomas derived from lower cranial nerve (IX, X, and XI) account for approximately 3% of all intracranial schwannomas in the absence of neurofibromatosis (6). These tumors generally occupy the jugular foramen and may also invade the surrounding bone, leading them to be commonly classified as jugular foramen schwannomas (JFSs). Researchers have

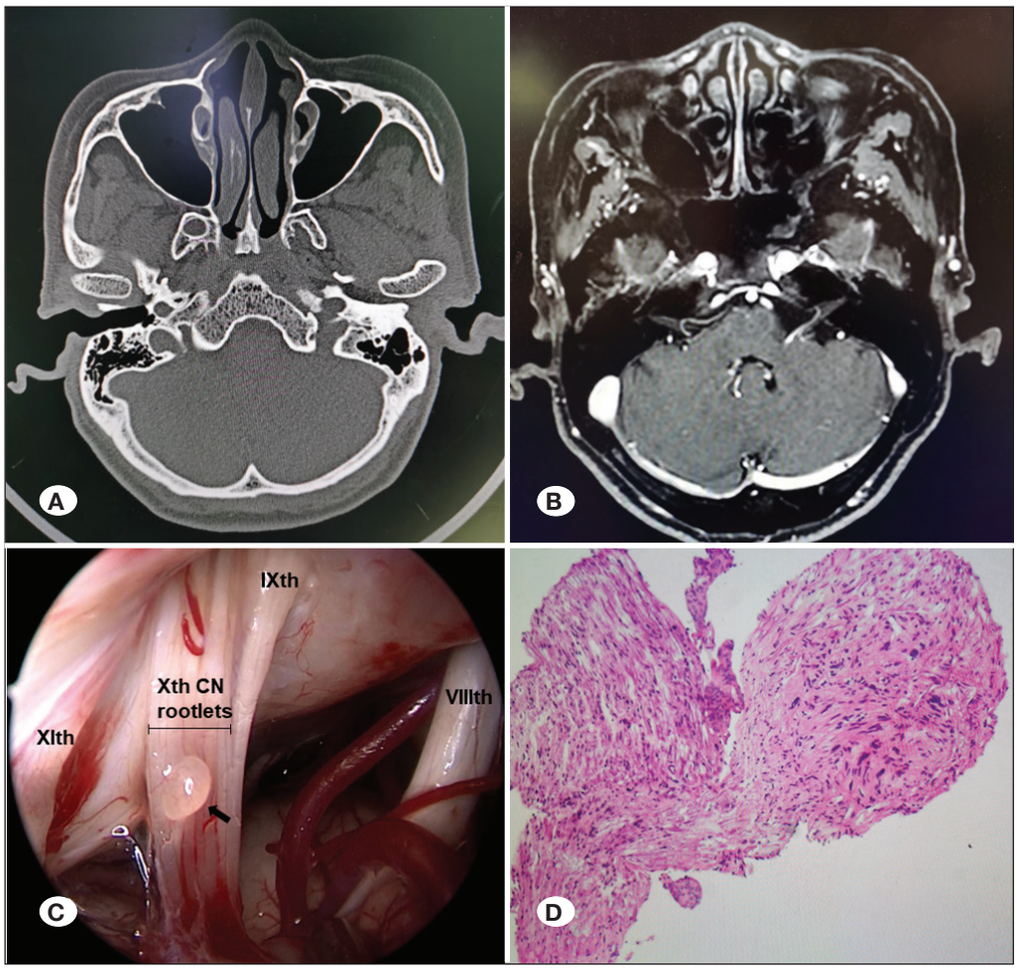

Figure 1: Preoperative CT scan indicated normal anatomy of the jugular foramen **(A)**; contrast-enhanced MRI revealed a point of contact between the left facial nerve and a small artery **(B)**; during endoscopic surgery, a neoplasm originating from the vagus nerve on the second rootlet from the caudal side was incidentally found **(C)**; postoperative pathology confirmed a benign schwannoma **(D)**.

Table I: Review of Previously Reported Cases of Purely Intracranial Vagal Schwannoma

Reference	Gender	Age	Neurofibromatosis	Initial symptoms	Tumor texture	Tumor origin	Extent of resection	Intraoperative condition	Postoperative complications
Sawamura and de Tribolet 1990 (16)	Female	36	No	syncopal attacks and postural hypotension	solid	NA	totally	NA	temporary hoarseness and dysphagia
Kawamura and See 1992 (8)	Female	72	NA	hoarseness and headache	cystic	NA	totally	NA	NA
Kamiguchi et al. 1995 (7)	Male	55	NA	hearing disturbance and tinnitus	solid	rostral rootlet	totally	NA	mild swallowing difficulties and hoarseness
Saito et al. 2000 (13)	Female	64	NA	hearing loss and tinnitus	solid	rostral rootlet	totally	NA	NA
Sharma et al. 2001 (18)	Male	60	No	hoarseness and intractable tinnitus	solid-cystic	one rootlet	totally	bradycardia asystole	hoarseness
Flint et al. 2005 (4)	Female	60	No	ataxia, nausea, vomiting, and headaches (hydrocephalus)	solid-cystic	three rootlets	NA	uneventful	temporary dysphonia and dysphagia
Kamel et al. 2006 (6)	Male	65	No	refractory hypertension, ataxia	solid	rostral rootlet	totally	uneventful	cerebral infarction
Gazzeri et al. 2009 (5)	Male	28	No	refractory hypertension (hearing loss) hoarseness	solid-cystic	rostral rootlet	totally	uneventful	cerebrospinal fluid fistula, meningitis
Zhao 2010*	Female	57	No	Glossopharyngeal neuralgia	solid	the third rootlet	totally	bradycardia hypotension	No
Schwam et al. 2019 (17)	Female	59	No	vertigo, ataxia, and hearing loss	solid	NA	totally	uneventful	No
Yao 2024	Female	59	No	asymptomatic	solid	caudal rootlet	totally	uneventful	No

*****This reference was published full text in Chinese. **NA:** not available.*

proposed various classifications to guide surgical approaches and predict prognosis for JFSs (Table II). In the 1980s, Kaye and Pellet categorized JFSs into four types, and later, Samii proposed a similar classification (9,12,15). Over subsequent years, purely intracranial schwannomas were reported in sporadic cases, leading researchers to suggest the addition of an A1 subtype that specifically refers to purely intracranial schwannomas. Bulsara independently classified this subtype as type A (3) and applied similar grading standards for classification of hypoglossal schwannomas that shared an analogous tumor growing pattern (11). In a recent update of the grading scale, Samii also separately listed the purely intracranial schwannomas as class A (14).

For lower cranial nerve schwannomas, it was difficult to recognize a specific nerve or a specific segment of nerve from which tumors were derived, as they tended to grow into large masses and invaded surround tissues by the time they were clinically diagnosed (10). Precise differentiation of the nerve segment involved is only possible when tumors are small and diagnosed at an early stage. Given that superior ganglia of the vagus nerve are located in the jugular foramen, it was hypothesized that schwannomas are more likely to originate from Schwann cells within ganglion areas, which subsequently extended to intra- or extracranial regions (19). In our study, we could clearly determine that the tumor originated from Schwann cells of the cerebellomedullary cistern rather than ganglion areas. We believe that the difference between intracranial and extracranial schwannomas can be attributed to distinctive locations of tumor origin rather than opposite directions of tumor extension.

As reviewed above, only 11 cases of purely intracranial vagal schwannomas have been documented so far. Despite summarizing the available data, drawing definitive conclusions remains challenging due to the limited information. However, several key observations are worth noting.

First, a majority of these tumors were derived from the rostral rootlets of the vagus nerve. Only one tumor originated from the caudal rootlet; this patient did not manifest any symptoms. This distinction is crucial because caudal rootlets consist of parasympathetic fibers, while rostral rootlets primarily comprise motor and sensory fibers and are located closer to the VIII/IX cranial nerves and rostral ventrolateral medulla (RVLM). This anatomic feature predisposes lesions present on the rostral rootlets to induce early-phase clinical symptoms such as hoarseness, tinnitus, and hypertension (6,8,18). It is speculated that the asymptomatic tumors were prone to derive from the caudal rootlets, which are situated farther from areas where compression causes significant symptoms.

Second, concerning surgical intervention and prognosis, although tumors varied in size, nearly all the tumors were totally resected using a retrosigmoid approach, and the patients achieved favorable prognosis owing to the confined growth of tumors without erosion of the jugular foramen. This is in line with a point mentioned previously that the choice of surgical approach and prognosis mainly hinges on the location of foramen schwannomas (2,3).

Table II: Classifications of Jugular Foremen Schwannoma

Classification	Kaye and Pellet	Samii	Bulsara	Samii
A	cerebellopontine angle with minimum enlargement of jugular foramen and with a small extension into the bone	cerebellopontine angle with minimum enlargement of jugular foramen	purely intradural tumors	tumor arising from cisternal part of the nerves, without significant extension into the JF
B	invading the bone (jugular foramen) with or without an intra-dural component	jugular foramen with intracranial extension	intracranial tumors that extend into the JF canal	intraosseous tumor inside the JF, extending into cisternal space or infratemporal fossa
C	extra cranial in location with a minor extension to the bone	extracranial tumor with extension into jugular foramen	dumbbell-shaped tumors with intracranial, intraosseous, and extracranial components	tumor arising from the peripheral part of the nerve
D	saddle bag shaped tumor with intra cranial and extracranial components	dumb bell-shaped tumor with both intra and extracranial components		triple dumbbell-shaped tumor with intracranial, intraosseous and extracranial parts

JF: *jugular foremen.*

Third, during two operation procedures, serious bradycardia and even asystole occurred, which were resolved with timely drug interventions. These occurrences were probably due to rapid decompression of the vagus nerve and subsequent rebound effect of vagal hyperactivity. Consequently, it is imperative for surgeons to be particularly vigilant regarding potential incidence of significant cardiovascular complications during such surgical interventions.

CONCLUSION

Purely intracranial vagal schwannoma is considered to be a rare subtype of schwannomas. In this report, we present a unique case in its asymptomatic early phase, definitively delineating its point of origin and precise anatomical location. A comprehensive review of the existing literature plays a pivotal role in enhancing our knowledge on tumor classification, clinical attributes, and prognosis.

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Ethical Approval

The study was conducted in accordance with the Declaration of Helsinki. All information is de-identified, including the photograph and radiological images.

Consent for Publication

Written informed consent has been obtained from the patient to publish this paper.

AUTHORSHIP CONTRIBUTION

Study conception and design: ZY
Data collection: ZY
Analysis and interpretation of results: ZY, SX
Draft manuscript preparation: ZY
Critical revision of the article: SX
Other (study supervision, fundings, materials, etc...): SX
All authors (ZY, SX) reviewed the results and approved the final version of the manuscript.

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