

---

# Spinal Accessory Nerve Schwannoma Involving the Jugular Foramen

O. Ortiz and L. Reed

**Summary:** We describe a patient with a schwannoma involving the intracranial, foraminal, and extracranial portions of the spinal accessory nerve.

**Index terms:** Neuroma; Nerves, accessory (XI); Foramina, jugular

Cranial nerve schwannomas are not uncommon intracranial tumors. Schwannomas arising from cranial nerve XI not only comprise a minority of these neoplasms but may also involve the extracranial segment of the spinal accessory nerve. The clinical presentation of cranial nerve XI schwannomas relates to their location and extent: intracranial, jugular foramen, upper neck, or cervical spine. This report describes the imaging features of a multilevel cranial nerve XI schwannoma.

## Case Report

A 76-year-old man presented with a 6-week history of headaches, nausea, and vomiting. The patient also had increasing unsteadiness on his feet. His medical history was remarkable for stage IB prostate cancer, treated with local excision and radiation therapy. Physical examination demonstrated atrophy of the left sternocleidomastoid and trapezius muscles. The upper extremity motor and sensory examinations as well as the remainder of the lower cranial nerve examination were unremarkable.

Cranial magnetic resonance (MR) imaging with spin-echo technique revealed a left jugular foramen mass with both intracranial and extracranial components. The lesion was isointense to brain parenchyma on T1-weighted (500/30/1 [repetition time/echo time/excitations]) images and predominantly hyperintense on T2-weighted (2500/80/2) sequences (Fig 1A–C). Prominent enhancement was noted after the intravenous administration of gadopentetate dimeglumine, except for a central nonenhancing region within the intracranial component, which presumably represented necrosis (Fig 1D). The lesion exerted mass effect on the medulla. The left trapezius and sternocleidomastoid muscles were atrophic (Fig 1C). High-resolution computed tomography (CT) of the temporal bone

was performed then (Fig 1E); transaxial images revealed relatively smooth expansion of the left jugular foramen. Calcifications were not present within the mass. Cerebral angiography demonstrated anteromedial displacement of the left cervical internal carotid artery without evidence of encasement (Fig 1F). A vascular stain was not identified on selective carotid or vertebral injection.

The intracranial portion of the tumor was debulked through a left retrosigmoid occipital craniectomy. The tumor encased cranial nerve XI, but displaced the remainder of the lower cranial nerves. The gross specimen consisted of a discrete mass of tan and yellow soft tissue with focal maroon areas (Fig 1G). Microscopically the lesion was composed of fascicles of polygonal, stellate, and fusiform cells with indistinct cytoplasmic margins. In some areas their nuclei were arranged in register-forming ill-defined palisades. The neoplasm was traversed by numerous blood vessels with thick, hyalinized walls. Cystic spaces occasionally were present as well as foci of lipid-laden and hemosiderin-laden macrophages (Fig 1H).

## Discussion

### *Schwannoma of the Spinal Accessory Nerve*

Although not an uncommon head and neck tumor, schwannomas that arise within the jugular foramen are rare. A review of the world literature in 1979 yielded 42 cumulative cases; scattered case reports are subsequently encountered (1).

In patients without phakomatosis, these lesions most often are encountered in the third to sixth decades of life. These tumors are slightly more common in female subjects (1). The clinical presentation of a patient with a jugular foramen schwannoma is quite variable. Symptoms may be ascribed to the initial lower cranial nerve of origin with variable onset and degree of compromise. Additionally, two or more lower cranial nerves may be affected, resulting in the classic jugular foramen syndrome (loss of taste in the posterior third of the tongue, vocal cord

---

Received September 13, 1993; accepted after revision January 5, 1994.

From the Departments of Radiology (O.O.) and Pathology (L.R.), West Virginia University, Robert C. Byrd Health Sciences Center, Morgantown.

Address reprint requests to O. Ortiz, Department of Radiology, Robert C. Byrd Health Sciences Center, PO Box 9235, Morgantown, WV 26506-9235.

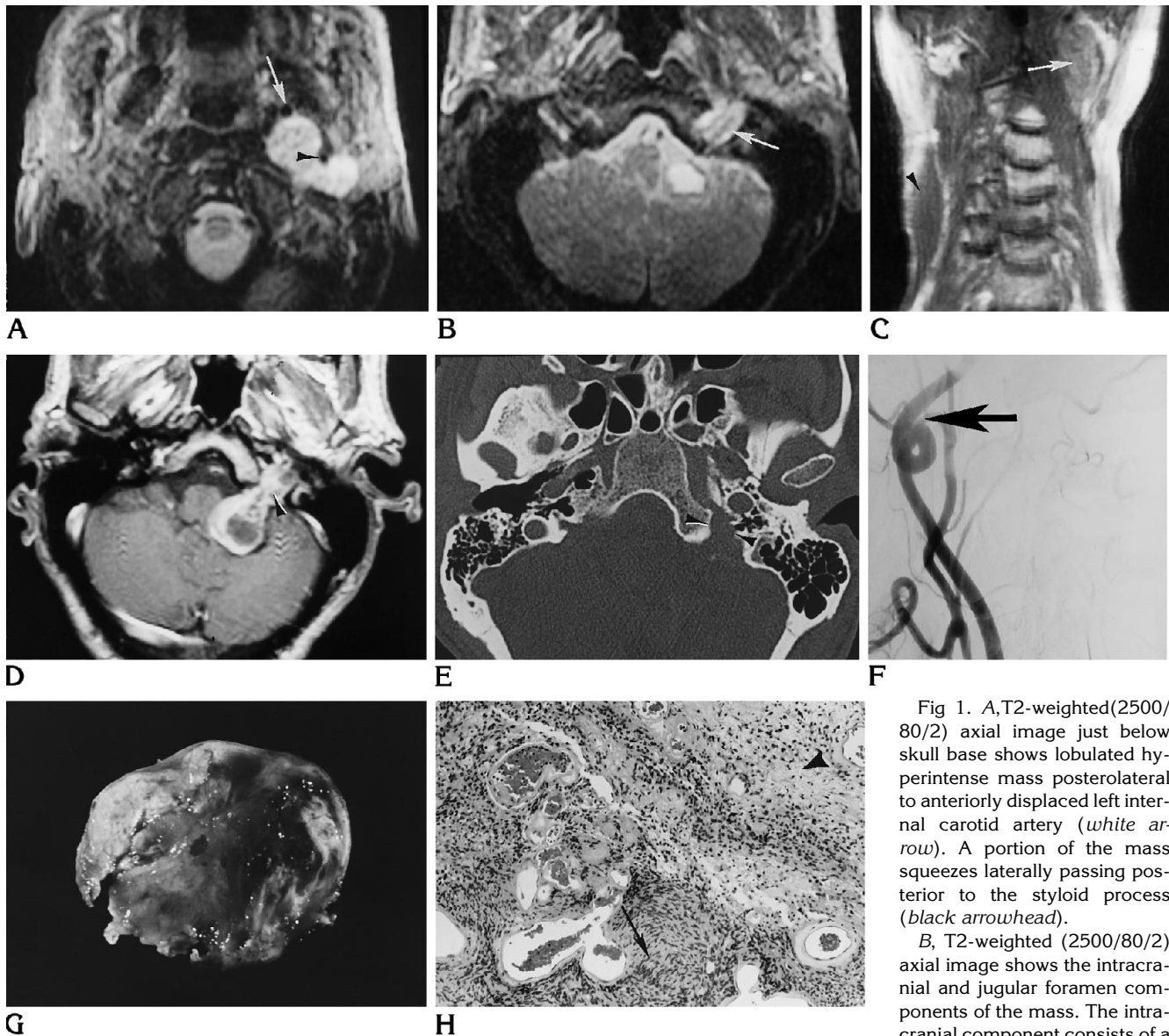


Fig 1. A, T2-weighted (2500/80/2) axial image just below skull base shows lobulated hyperintense mass posterolateral to anteriorly displaced left internal carotid artery (*white arrow*). A portion of the mass squeezes laterally passing posterior to the styloid process (*black arrowhead*).

B, T2-weighted (2500/80/2) axial image shows the intracranial and jugular foramen components of the mass. The intracranial component consists of a

large central hyperintense area surrounded by an intermediate-intensity rim. Mass effect on the dorsolateral aspect of the medulla is noted. The foraminal component is heterogeneously hyperintense and expands the jugular foramen (*white arrow*).

C, T1-weighted (500/30/1) coronal image demonstrates an oval-shaped, well-circumscribed mass (*arrow*) with intermediate signal intensity extending from the skull base into the superior carotid space. The left sternocleidomastoid muscle is atrophic. Sternocleidomastoid on normal side is indicated by *black arrowhead*.

D, Postcontrast T1-weighted (500/30/1) axial image reveals uniform enhancement of the solid portions of the tumor and lack of enhancement within the center of the intracranial component. The tumor can be followed into the jugular foramen (*arrowhead*).

E, Axial CT with bone algorithm shows smooth expansion of the left jugular foramen (*arrowheads*).

F, Selective left common carotid artery injection demonstrates absence of neovascularity and tumor stain. Avascular mass effect with anterior displacement (*arrow*) of the distal cervical internal carotid artery is noted.

G, Grossly, the neoplasm forms a discrete, encapsulated soft mass. The cut surface demonstrated color variation ranging from tan to yellow. Areas of old hemorrhage are evident.

H, Microscopically the neoplasm is composed of areas of fusiform cells arranged in compact fascicles (Antoni A pattern) (*arrow*) alternating with looser areas (Antoni B pattern) (*arrowhead*).

Cranial nerve XI schwannomas

Author (Year)	Reference	Patient Age, y/Sex	Tumor Location
Ruberti and Carteri (1961)	(1)	31/M	JF
Pluchino et al (1975)	(1)	42/F	JF
Pou-Serradell (1978)	(8)	56/F	JF
Christoferson (1982)	(9)	24/F	C
Tsuchiya et al (1982)	(6)	18/M	C
Tohyama et al (1982)	(6)	52/M	C, JF, EC (C-2 level)
Julow (1983)	(6)	50/F	C
Matsushima et al (1985)	(10)	51/M	JF
Kawaguchi et al (1987)	(6)	38/F	EC (C-2 level)
Nakashima et al (1988)	(2)	67/M	JF
Lanzieri (1990)	(11)	...	C, JF, EC
Present case (1992)		76 /M	C, JF, EC

Note.—C indicates cisternal; JF, jugular foramen; and EC, extracranial.

paralysis, dysphagia, weakness of the sternocleidomastoid and trapezius muscles) or its variants. Patients also may present with signs and symptoms of cerebellar involvement and/or increased intracranial pressure and nystagmus (1).

Schwannomas of cranial nerve XI represent an even rarer form of jugular foramen mass (Table). These tumors are defined as arising specifically from or encasing cranial nerve XI. The extent of cranial nerve involvement may include the cisternal, foraminal, and extracranial (spinal or intrasternomastoid) segments of the spinal accessory nerve. These patients usually present with a partial or complete jugular foramen syndrome (2). The characteristic physical examination finding is denervation atrophy of the trapezius and sternocleidomastoid muscles (3). Of the remaining lower cranial nerves, cranial nerve VIII is most often compromised, and cranial nerve XII is infrequently affected. Hypoglossal nerve dysfunction tends to occur with large intraforaminal masses. When a prominent intracranial component of tumor is present, signs and symptoms referable to brain stem and cerebellar dysfunction may be identified.

### Pathology

Schwannomas characteristically arise focally from the sheath of a nerve fascicle forming a well-demarcated, eccentric mass that deflects

the parent nerve. The neoplasms are encapsulated with a smooth, sometimes lobulated, external surface. Secondary degenerative changes commonly are present in large lesions and include yellow fatty areas and cysts of varying sizes. Red or maroon discoloration may be present, signifying recent or remote hemorrhage.

Schwannomas are classically composed of two distinct alternating architectural patterns, designated *Antoni A* and *Antoni B* areas. *Antoni A* areas comprise compact strands and fascicles of spindle cells that are delicately fibrillated. In contrast, *Antoni B* areas consist of more loosely arranged vacuolated cells that tend to be pleomorphic.

### Imaging

Cranial nerve XI schwannomas are usually located within or involve the jugular foramen (Table). The jugular foramen may be expanded, a finding that has been reported not only with cross-sectional imaging but also with conventional skull radiography. Because it is a slowly growing mass, the margins of the jugular foramen are smoothly eroded. The intracranial and extracranial components of the tumor are well-circumscribed and lobulated, with associated mass effect on adjacent neural (brain stem, cerebellum, spinal cord) or soft-tissue structures of the head and neck regions, respectively.

The CT and MR appearance of cranial nerve XI schwannomas parallels that of cranial nerve VIII schwannomas. On CT, these tumors are hypointense or isodense relative to brain parenchyma. They may contain cystic or necrotic foci but do not manifest a calcified component. On conventional spin-echo MR, schwannomas are hypointense or isointense to brain on T1-weighted images and variably intense on proton-density and T2-weighted images (4). Areas of necrosis or cyst formation appear hypodense on CT, hypointense on T1-weighted spin-echo MR, and hyperintense on T2-weighted sequences. After intravenous contrast administration, these schwannomas will demonstrate moderate enhancement of solid components on both CT and MR. Early scans reveal slightly heterogeneous enhancement, whereas delayed scanning shows a more homogeneous pattern (5).

A combination of modalities including angiography and cross-sectional imaging may be

used to help distinguish intraforaminal cranial nerve XI schwannomas from other jugular foramen masses. With schwannomas, the arterial phase demonstrates vascular displacement and stretching; the distal cervical internal carotid artery or posterior inferior cerebellar artery may be involved. A mild to moderate tumor stain is infrequently present (6). Intraforaminal schwannomas are associated with absent venous filling of the involved jugular foramen. Paragangliomas are hypervascular lesions on angiography; they tend to be ill-defined on CT and may demonstrate a "salt and pepper" appearance on T2-weighted MR images (7). A metastasis will show minimal angiographic vascularity, unless secondary to a hypervascular primary tumor. The margins of a jugular foramen metastasis will be irregular. The MR signal characteristics of jugular foramen metastasis usually are similar to the signal characteristics of schwannomas in this location (7). Chondrosarcomas and meningiomas that involve the jugular foramen demonstrate minimal angiographic vascularity. Cross-sectional imaging may reveal calcific components. An asymmetrically enlarged jugular fossa initially may present as a "pseudomass." Careful scrutiny of precontrast and postcontrast CT and/or MR images usually will identify this normal variant.

Schwannomas of the lower cranial nerves account for only a small percentage of jugular foramen masses (7). Schwannomas of cranial nerve XI are even less common. Furthermore, it is often difficult to distinguish the nerve of origin of these masses by imaging and surgery, particularly when the lesion attains a large size and

involves multiple cranial nerves. The clinical presentation, the jugular foramen syndrome, likewise usually does not separate these tumors. Other tumors that must be included in the differential diagnosis of a jugular foramen mass include paraganglioma, metastasis, chondrosarcoma, and meningioma.

## References

1. Hakuba A, Hashi K, Fujitani K, Ikuno H, Nakamura T, Inoue Y. Jugular foramen neurinomas. *Surg Neurol* 1979;11:83-94
2. Nakashima T, Tomita K, Uemura T, Matsushima T, Fukui M. Accessory nerve schwannoma extending extracranially to the second cervical level of vertebra. *J Laryngol Otol* 1988;102:959-961
3. Noyek AM, Chapnik JS, Wortzman G, Kandel R. Schwannoma of the intrasternomastoid portion of the spinal accessory nerve: sophisticated pre-operative MRI diagnosis and appropriate surgical management. *J Otolaryngol* 1992;21:286-289
4. Bilaniuk LT. Adult infratentorial tumors. *Semin Roentgenol* 1990;25(2):155-173
5. Hasso AN, Smith DS. The cerebellopontine angle. *Semin Ultrasound, CT MR* 1989;10(3):280-301
6. Kawaguchi S, Ohnishi H, Yuasa T, Hashimoto H. Spinal accessory nerve neurinoma in the C2 spinal canal. *Neurologia Medico-Chirurgica* 1987;27:1190-1194
7. Lo WW. Tumors of the temporal bone and the cerebellopontine angle. In: Som PM, Bergeron RT, eds. *Head and Neck Imaging*. St Louis: Mosby; 1991:1046-1108
8. Pou-Serradell A, Llovet-Tapies J, Galito E, Pascual J. Neurinome intracranien du nerf spinal. *Rev Neurol (Paris)* 1978;134:803-805
9. Christoferson LA, Leech RW, Grossman M. Intracranial neurilemmoma of the spinal accessory nerve. *Surg Neurol* 1982;18:18-20
10. Matsushima T, Fukui M, Matsunaga M, Kitamura K, Hasuo K. Accessory nerve neurinoma mimicking a brain stem tumor on angiography: report of a case. *Neurosurgery* 1985;16:839-842
11. Lanzieri CF. MR imaging of the cranial nerves. *AJR Am J Roentgenol* 1990;154:1263-1267