The jugular foramen transmits vital neurogenic and vascular structures that are of immense importance during skull base surgery.21 These neurovascular structures may give rise to a variety of lesions, most commonly glomus jugulare tumors, meningiomas, and schwannomas.20 Cavernous hemangiomas are benign vascular malformations that commonly occur as cerebral intraaxial lesions. They also frequently arise extracranially in any vascularized tissue, including within the liver, skin, muscle, and bone, and occasionally they develop as multifocal lesions.29 The occurrence of cavernous hemangiomas in the jugular foramen, however, is extremely uncommon, with only one case previously reported.17 To the best of our knowledge, we report the first case of a multifocal cavernous hemangioma of the jugular foramen, which was treated with a radical excision via a right infratemporal fossa approach and high neck dissection.

Case Report
Clinical Presentation
A 60-year-old woman presented with a 9-month history of a right-sided neck mass. Although the mass remained stable in size, she developed significant right-sided suboccipital pain and neck discomfort while turning the head contralaterally. The patient also reported having progressive dysphagia and hoarseness over 3 months. She had no personal or family history of malignancy, radiation exposure, vascular malformations, or neurofibromatosis.

On examination, a soft, slightly tender, $3 \times 3$–cm mass
was noted in the upper third portion of the sternocleidomastoid muscle (SCM) on the patient’s right side. There was wasting of the right SCM and trapezius muscle, deviation of the uvula to the left, and decreased pharyngeal sensation on the right side, while the patient’s gag reflex was intact. The remaining clinical findings were unremarkable. After examination of two fine-needle aspirates from the primary neck mass did not provide a diagnosis, the patient underwent an open biopsy; the findings were consistent with a cavernous hemangioma.

Imaging Studies and Laboratory Evaluation

Brain gadolinium-enhanced MRI and brain and neck contrast-enhanced CT scanning depicted a heterogeneously enhancing, right-sided lesion expanding the right jugular foramen and compressing the jugular bulb. The lesion had a small intracranial component, displaced the facial nerve in the fallopian canal laterally, and abutted the vertical segment of the petrous carotid artery. The lesion additionally appeared to involve the anterior and inferior parts of the right occipital condyle and the ipsilateral anterior arch of C1, and it extended into the neck under the right-sided SCM in a multifocal fashion (Fig. 1). Notably, digital subtraction angiography performed preoperatively demonstrated a patent but significantly stenosed jugular bulb with dominant contralateral venous outflow.

FIG. 1. Preoperative T2-weighted MR images demonstrating a hyperintense lesion in the region of the jugular foramen as well as several vertically aligned lesions in the neck under the SCM (A: coronal; C and F: axial). Postoperative T1-weighted (B: coronal; E and H: axial) and T2-weighted (D and G: axial) MR images demonstrating gross-total resection of the masses in the jugular foramen and in the neck.

110   J Neurosurg Volume 131 • July 2019
Operation

The patient was positioned supine, and a right-sided C-shaped incision was made 4.5 cm posterior to the postauricular crease. The incision was extended 2 fingerbreadths below the inferior border of the mandible, allowing the neck dissection to be performed. After the tail of the parotid gland was elevated off the anterior border of the SCM, a T-shaped incision was made at the temporal line and the anterior border of the SCM. A temporalis fascial flap was elevated, and it was left pedicled to the anterior branch of the superficial temporal artery. The mastoid was exposed, and the SCM and posterior digastic attachments to the mastoid tip were removed. The styloid process was then identified superiorly, and the tip was removed. The dissection continued inferiorly until the omohoid muscle was identified, marking the inferior border of the exposure. The internal carotid artery, internal jugular vein, and cranial nerves IX, X, XI, and XII were each dissected and exposed up to the skull base. A large nodal mass and multiple smaller subscalene masses were resected. Finally, having confirmed adequate contralateral venous outflow on preoperative angiography, we suture ligated the internal jugular vein distal to the inferior extension of the mass.

Attention was then turned to the mastoid, where a standard mastoidectomy was performed with a high-speed drill and the sigmoid sinus exposed. The facial recess was opened and the incudostapedial joint divided. The facial nerve was then skeletonized to the stylomastoid foramen. After additional drilling of the osseous external auditory canal and removal of the mastoid tip, the retrofacial air cells were removed and the jugular bulb exposed. The facial nerve was then transposed and held in place with a cuff of muscle in a tensionless fashion. At this point, the jugular vein was brought through the foramen. After occlusion of the sigmoid sinus with Surgicel, the tumor was progressively debulked to expose the presigmoid dura. The posterior fossa dura was opened, allowing the identification of the right vertebral artery as well as cranial nerves IX, X, and XI. There was no evidence of intradural extension of the right vertebral artery. 21

Attention was then turned to the mastoid, where a standard mastoidectomy was performed with a high-speed drill and the sigmoid sinus exposed. The facial recess was opened and the incudostapedial joint divided. The facial nerve was then skeletonized to the stylomastoid foramen. After additional drilling of the osseous external auditory canal and removal of the mastoid tip, the retrofacial air cells were removed and the jugular bulb exposed. The facial nerve was then transposed and held in place with a cuff of muscle in a tensionless fashion. At this point, the jugular vein was brought through the foramen. After occlusion of the sigmoid sinus with Surgicel, the tumor was progressively debulked to expose the presigmoid dura. The posterior fossa dura was opened, allowing the identification of the right vertebral artery as well as cranial nerves IX, X, and XI. There was no evidence of intradural extension of the mass.

At the completion of tumor removal, abdominal fascia was grafted to patch the dural opening. The facial nerve was repositioned, the eustachian tube was plugged, and the mastoid defect was packed with abdominal fat. The previously harvested temporoparietal facial flap was laid over the mastoid tip with the posterior margin that encompasses vital neurovascular structures. 21 Not uncommonly, a tumor can arise within the jugular foramen and impinge on adjacent neurovascular structures, giving rise to lower cranial nerve neuropathies. Known as jugular foramen syndrome, these neuropathies typically involve cranial nerves IX, X, and XI, although 30% of the cases additionally involve the hypoglossal nerve. 4,7,20 Glomus jugulare paragangliomas are the most commonly encountered tumors within the jugular foramen. 20 Other less frequently encountered lesions include meningiomas, schwannomas, aneurysmal bone cysts, cholesteatomas, chondromas, and inflammatory granulomas. 20 Radiological imaging of these lesions not uncommonly overlaps. 12,20 Thus, definitive diagnosis often requires formal histopathological examination.

Discussion

The jugular foramen is an important anatomical landmark that encompasses vital neurovascular structures. 21 Not uncommonly, a tumor can arise within the jugular foramen and impinge on adjacent neurovascular structures, giving rise to lower cranial nerve neuropathies. Known as jugular foramen syndrome, these neuropathies typically involve cranial nerves IX, X, and XI, although 30% of the cases additionally involve the hypoglossal nerve. 4,7,20 Glomus jugulare paragangliomas are the most commonly encountered tumors within the jugular foramen. 20 Other less frequently encountered lesions include meningiomas, schwannomas, aneurysmal bone cysts, cholesteatomas, chondromas, and inflammatory granulomas. 20 Radiological imaging of these lesions not uncommonly overlaps. 12,20 Thus, definitive diagnosis often requires formal histopathological examination.

Hemangiomas are classified into 3 distinct categories: capillary, cavernous, and mixed. The former usually occurs in children and obliterates spontaneously, 24,26 while the latter two pathological subtypes tend to persist, becoming symptomatic via hemorrhage or mass effect on surrounding structures. 24,26 Cavernous hemangiomas constitute 5%–13% of all vascular lesions in the brain and spinal cord. 15,23 They are endothelial-lined, grossly dilated (cavernous), thin-walled vascular malformations that lack the
Cavernous hemangiomas most commonly arise in the central nervous system parenchyma. Less frequently, they have been reported in the tentorium, skull bones, skull base, infratemporal fossa, anterior jugular vein, carotid sheath, petrosal sinus, falk cerebri and convexity dura, cerebellopontine angle, Meckel’s cave, and cranial nerves. Notably, Menzel and Denecke described the first and only case to date of a cavernous angioma of the jugular foramen in a 51-year-old woman who presented with jugular foramen syndrome. As in the current patient, imaging demonstrated enlargement of the right jugular foramen with compression of the jugular bulb. Surgical extirpation was achieved via a radical mastoidectomy with concurrent high neck dissection. At the 6-month follow-up, the authors reported complete functional recovery of cranial nerves IX and X with residual accessory nerve paresis.

Surgical resection of symptomatic cavernous hemangiomas remains the primary management option. In quiescent lesions, where impingement of adjacent neurovascular structures is often absent, a conservative approach with observation and radiological surveillance is acceptable, particularly when aggressive therapy of the lesion may result in untoward neurological sequelae. The role of radiation therapy in the management of cavernous hemangiomas is controversial. The current literature lacks evidence-based results regarding appropriate application and therapeutic efficacy among these patients. Moreover, while radiosurgical treatment of cavernous hemangiomas located in high-risk eloquent areas has been shown to reduce rehemorrhage rates, there are no data demonstrating significant lesion regression or improvement in mass effect related to the use of radiation among symptomatic patients.

To the best of our knowledge, there have been no previous reports of a multifocal cavernous hemangioma involving the jugular foramen. Similar to the case presented by Menzel and Denecke, our patient had an extremely rare presentation of jugular foramen syndrome secondary to a cavernoma. While the lesions in both cases enlarged the jugular foramen and obstructed flow in the jugular bulb, in the current case, the lesion was multifocal in nature with extensive extracranial involvement. Surgical extirpation, with attempts to limit consequent cranial neuropathies, is recommended.

Acknowledgments

We would like to thank Kristin Kraus, MSc, for her contribution to manuscript editing and Vance Mortimer for his contribution to preparing the video.

References


Unauthenticated | Downloaded 01/19/24 09:03 PM UTC
Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Couldwell, Abou-Al-Shaar, Gozal. Acquisition of data: Abou-Al-Shaar, Gozal. Analysis and interpretation of data: all authors. Drafting the article: Abou-Al-Shaar, Gozal. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Couldwell.

Supplemental Information
Videos

Correspondence
William T. Couldwell: Clinical Neurosciences Center, University of Utah, Salt Lake City, UT. neuropub@hsc.utah.edu.