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Primary jugular foramen fibrous dysplasia: surgically nuanced video of extradural infratemporal transjugular approach. Illustrative case

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BACKGROUND Fibrous dysplasia of the bone is a disease caused by a somatic *GNAS* mutation that affects craniofacial bones and can have a mass effect on different neurovascular structures. The authors present the first case of primary jugular foramen fibrous dysplasia with occlusion of the transverse and sigmoid sinuses.

OBJECTIVE A 33-year-old man presented with a history of dizziness and occasional dysphagia over the past year. Magnetic resonance imaging showed a uniform enhanced mass in the left jugular foramen, with complete blockage of the adjacent transverse and sigmoid sinuses. The computed tomography scan revealed a cystic bone lesion of the jugular foramen. The patient underwent a gross-total removal of the tumor through an infratemporal transjugular approach with complete preservation of the lower cranial nerves.

LESSONS This first reported case of primary jugular foramen fibrous dysplasia highlights the importance of considering this diagnosis when evaluating jugular fossa lesions. Understanding the anatomy of the infratemporal and jugular fossae, along with proficiency in microsurgical techniques, is essential for removing such tumors while preserving cranial nerve functions and the patient's quality of life.

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KEYWORDS jugular fossa; jugular foramen; infratemporal approach; fibrous dysplasia; skull base; craniocervical junction

Fibrous dysplasia of the bone is a stem cell bone disease caused by a somatic *GNAS* mutation.[1] This condition is characterized by the slow, progressive replacement of a localized area of the bone by an abnormal proliferation of fibrous tissue mixed with poorly formed, disorderly arranged trabeculae of the woven bone.[2,3] Although craniofacial fibrous dysplasia is a nonmalignant and progressive disorder, it can cause a mass effect on the cranial structures.

In this report, we present the case of a healthy young male who experienced nonspecific dizziness, followed by choking and the discovery of a mass in the left jugular foramen, leading to complete blockage of the adjacent transverse and sigmoid sinuses. The patient underwent successful total removal of the tumor through an infratemporal posterior fossa transjugular approach, with preservation of full neurological function. Fibrous dysplasia involving the jugular foramen is described in patients as an extensive temporal and occipital disease. Here, we report a case of localized fibrous dysplasia of the jugular foramen in the occipital bone and present a nuanced video of the surgical approach.

Illustrative Case

A 33-year-old male with no prior medical history presented with symptoms of dizziness and occasional dysphagia over the last 12 months. He was referred to our department after magnetic resonance imaging (MRI) revealed a homogeneous enhanced lesion in the left jugular foramen (Fig. 1A). Magnetic resonance angiography (MRA) showed total occlusion of the left transverse and sigmoid sinuses (Fig. 1B). A computed tomography (CT) scan revealed an erosive cystic lesion around the jugular foramen, exclusively in the occipital bone (Fig. 1C). The preoperative neurological examination was normal.

The patient underwent surgery through an infratemporal posterior fossa transjugular approach.[4,5] The anatomical basis of the approach

ABBREVIATIONS CN = cranial nerve; CT = computed tomography; IJV = internal jugular vein; MRA = magnetic resonance angiography; MRI = magnetic resonance imaging; SCM = sternocleidomastoid.

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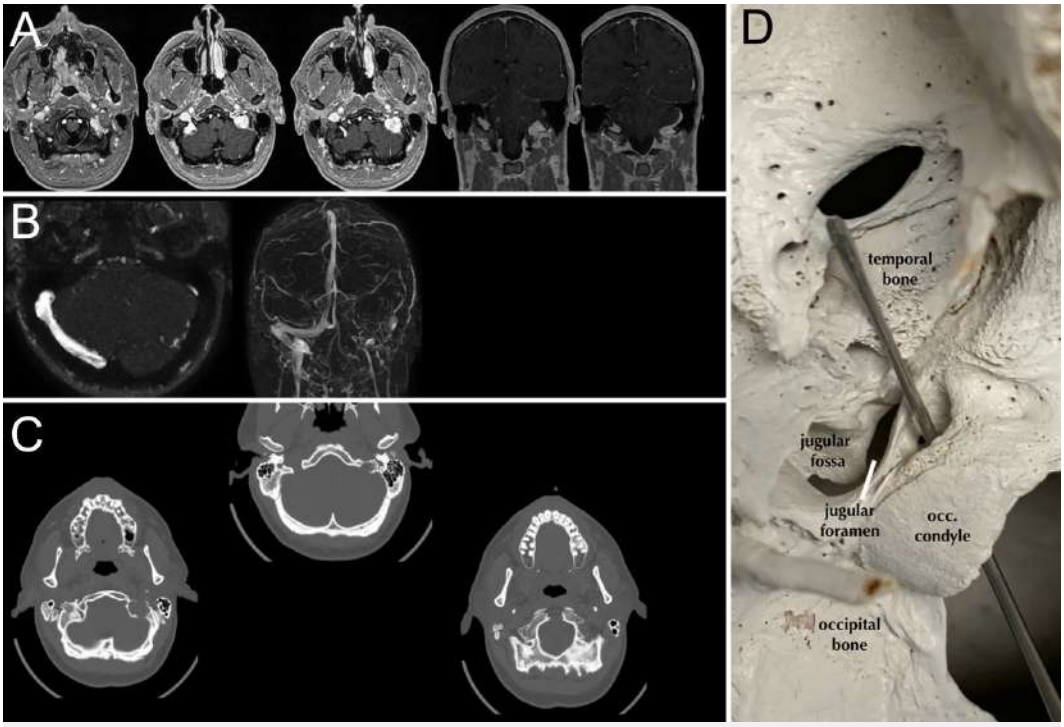

FIG. 1. MRI (**A**) and MRA (**B**) show a lesion with uniform contrast enhancement in the left jugular foramen and complete blockage of the left transverse and sigmoid sinuses. CT scans (**C**) showing a cystic erosive lesion of the occipital bone, compromising the jugular foramen and posterior part of the condyle without involvement of the petrous bone. Inferior view (**D**) of the skull base showing the jugular fossa and jugular foramen. occ. = occipital.

is illustrated in Fig. 2. The patient was positioned supine with the head turned 60° to the right side and extended. An arciform skin incision was made, starting 2 cm above the pinna and curving around 2 cm posterior to the asterion, and extending 4 cm below the mastoid tip, passing over the anterior margin of the sternocleidomastoid (SCM) muscle. During the dissection of the muscle and cervical region, we proceeded with layer-by-layer exposure. We identified the SCM muscle, cranial nerve (CN) XI, the internal jugular vein (IJV), the common carotid artery, internal and external carotid arteries, and CN XII. The vagus nerve, CN X, is located between the IJV and the carotid artery. In the sequence, we identified the splenius capitis muscle, the levator scapulae muscle, the digastric muscle, and the suboccipital triangle. Additionally, we located the obliquus capitis superior muscle and the obliquus capitis inferior muscle. A mastoidectomy and a retrolabyrinthine approach were performed, exposing the entire sigmoid sinus, including the jugular bulb. The tumor was located in the occipital bone, obstructing the jugular foramen, and infiltrating the occipital condyle. We completely removed the lesion and performed drilling on the occipital bone, opening the jugular foramen and posterior part of the occipital condyle (Video 1).

VIDEO 1. Clip showing the surgically nuanced video of the extradural infratemporal transjugular approach for a primary fibrous dysplasia of the left jugular foramen. Occ Cond = occipital condyle. Click here to view.

The postoperative CT scanning revealed complete bone removal around the lesion, and MRI showed no residual tumor (Fig. 3). The patient was discharged 4 days postsurgery with no neurological deficits. Histological study revealed fibrous dysplasia (Fig. 4).

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

Observations

Fibrous dysplasia of the bone is a well-described condition in which normal bone is replaced by abnormal fibroconnective tissue proliferation.[1] The clinical spectrum includes various forms, such as the most common monostotic form (70%), the polyostotic form (30%), the rare variant known as McCune-Albright syndrome, which is characterized by café-au-lait cutaneous spots and endocrine abnormalities, and the craniofacial forms.[2,3] According to various studies, the occipital bone is rarely involved in monostotic fibrous dysplasia.[3,6–8]

The management of fibrous dysplasia is a topic of debate. Patients experiencing bone pain can be treated with bisphosphonates, which are drugs that prevent osteoclastic bone resorption.[9] Surgery may be necessary for cases involving neurological complications.[10–12]

The definition of jugular fossa and its differentiation from the jugular foramen was clearly presented by Williams and Warwick[13] and Arnautović and Al-Mefty.[4] The jugular foramen is an opening in the skull that connects the posterior cranial fossa and the jugular fossa. The jugular fossa is a deep depression located at the bottom of the petrous part of the temporal bone. Its size can vary in different skulls. This depression connects to the posterior cranial fossa through the

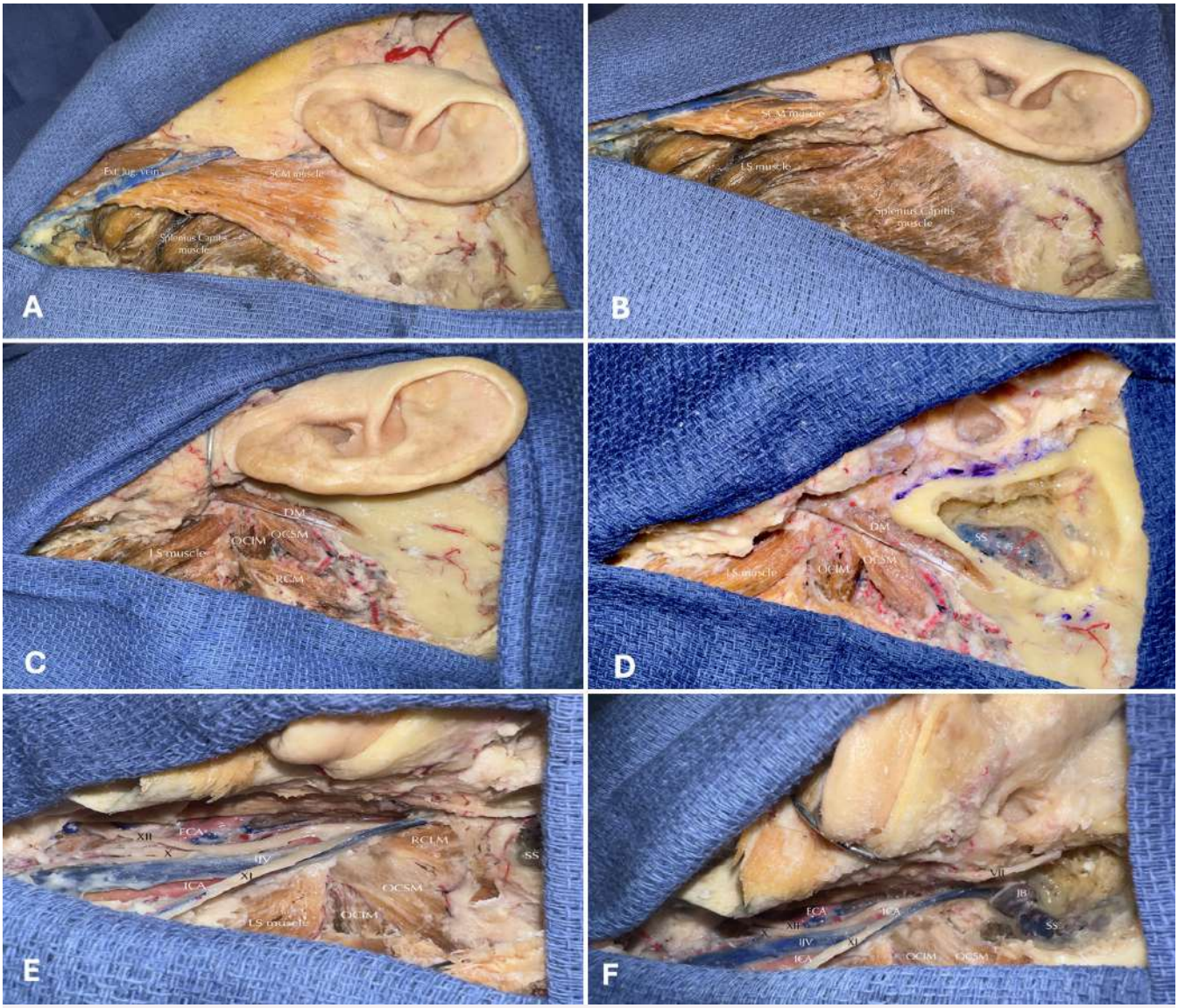

FIG. 2. Anatomical laboratory dissection (**A–F**). DM = digastric muscle; ECA = external carotid artery; Ext. Jug. = external jugular; ICA = internal carotid artery; JB = jugular bulb; LS = levator scapulae; OCIM = obliquus capitis inferior muscle; OCSM = obliquus capitis superior muscle; RCLM = rectus capitis lateralis muscle; RCM = rectus capitis major muscle; SS = sigmoid sinus; VII = CN VII; X = CN X; XI = CN XI; XII = CN XII.

jugular foramen and houses the jugular bulb, which continues as the jugular vein downward.[13] In the current case, as illustrated in Fig. 1, the lesion was found exclusively in the occipital bone, blocking the jugular foramen.

Radiologically, Fries categorized fibrous dysplasia of the cranium into 3 subtypes. The most common is pagetoid (56%), followed by sclerotic (23%) and cystic (21%) forms.[14] In our case, the predominant form was the rare cystic type, as depicted in Fig. 1. The signal on T1-weighted MRI studies can range from low to intermediate, depending on the ratio of fibrous tissue to mineralized matrix. Areas with more fibrous tissue tend to have an intermediate signal, while areas with a more mineralized matrix display a lower signal. The signal on T2-weighted images also varies, with predominantly fibrous areas showing high signal and low signal indicating regions with a more mineralized matrix. The fibrous component of fibrous dysplasia is well vascularized with small central blood vessels and peripheral sinusoids. As a result, fibrous dysplasia typically shows intense enhancement with gadolinium, as seen in the current case.[15] Table 1 presents the differential diagnosis of the jugular foramen lesions.

The extradural infratemporal transjugular approach is an excellent option for treating such tumors, as it provides superior exposure of the lesion's boundaries compared to the lateral suboccipital and traditional retrosigmoid approaches. The extended cervical dissection and exposure of the sigmoid sinus ensure safe management of the jugular bulb region and the control of bleeding from the inferior petrosal sinus. The dissection of the 9th, 10th, and 11th CNs is essential to identify and preserve their anatomical and functional integrity in the jugular foramen area. The condyle was drilled in its posterior third without risking craniocervical instability.

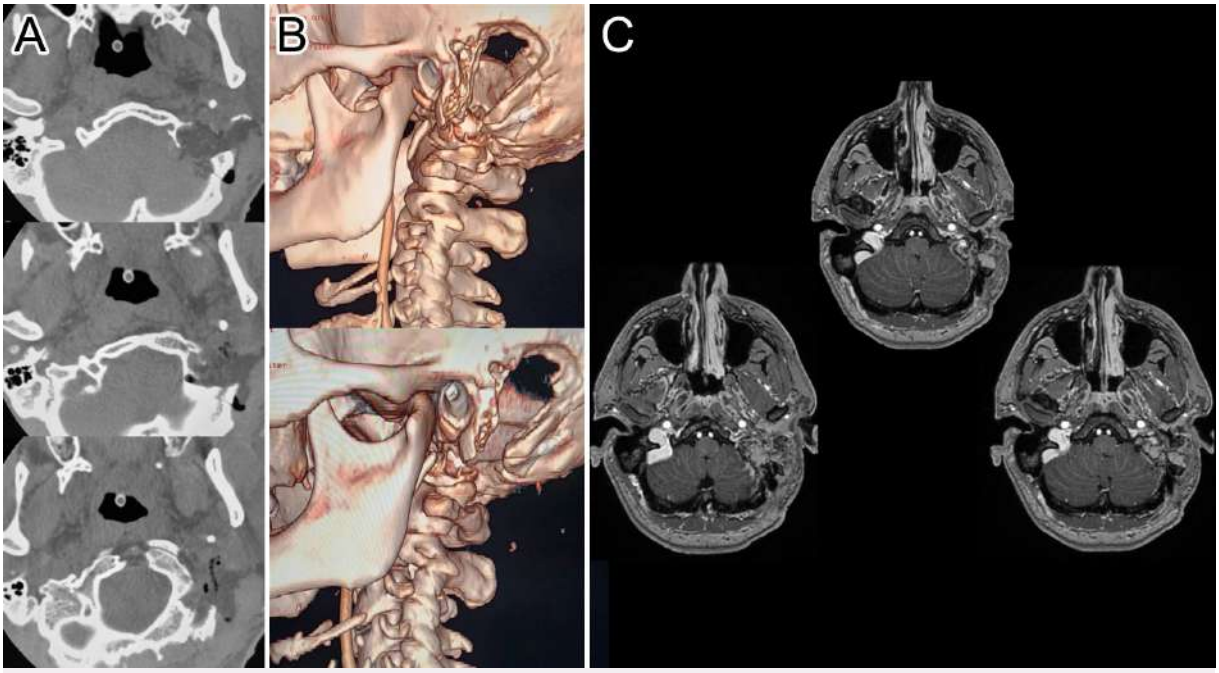

FIG. 3. Postoperative CT scans (**A**) and MRI studies (**C**). Axial images display complete bone removal of the cystic erosive occipital lesion, which has invaded the left jugular foramen. A three-dimensional bone reconstruction (**B**) demonstrates the lateral craniocervical junction bone resection.

We presented the first case of primary fibrous dysplasia related to the occipital bone in the jugular foramen, emphasizing the importance of considering this diagnosis when planning the surgical treatment of jugular fossa lesions. When surgery is necessary, complete removal is the most effective treatment for fibrous dysplasia.

Lessons

The surgical techniques for effectively treating jugular fossa tumors have been developed at several centers over the last few decades.[4,5,16–20] Systematic training in skull base surgery and step-by-step reproduction in the laboratory ensure the safe removal of jugular fossa tumors while preserving neurological function and quality of life.

Complete removal of the tumor without causing additional venous drainage damage was possible due to the total occlusion of the transverse and sigmoid sinuses, which had been caused by blockage of the jugular foramen by tumor growth. The tumor removal and preservation of the anterior part of the jugular bulb and IJV, as described by Al-Mefty and Teixeira,[16] were crucial for maintaining the normal function of the lower CNs.

The infratemporal posterior fossa transjugular approach provides complete exposure of the jugular fossa region and can extend to the condylar, mastoid, and petrosal areas. It is an excellent approach for treating complex lesions of the craniocervical junction. In this particular case, the comprehensive exposure allowed for complete removal of the tumor, which was the best option for the patient.

Surgery was necessary to prevent the invasion of the jugular fossa and the petrous part of the temporal bone, which could cause additional neurological deficits in the patient.

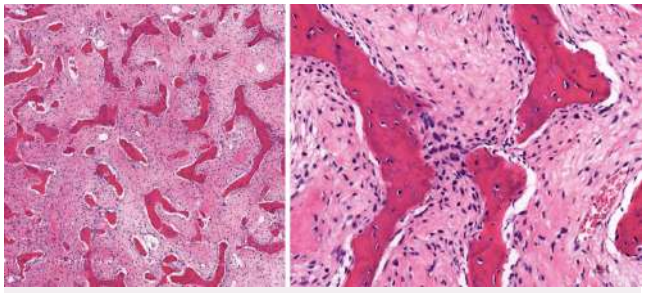

FIG. 4. Histological diagnosis of fibrous dysplasia. Hematoxylin and eosin, original magnification ×10 (**left**) and ×40 (**right**).

TABLE 1. Differential diagnosis of lesions in the jugular fossa

Tumor	CT Scan	MRI
Paraganglioma	Avidly enhances w/ contrast w/ delayed washout (due to rich capillary network)	"Salt & pepper," T1 inhomogeneous enhancement
Neuroma	"Bone scalloping" appearance & enlargement of jugular foramen	"Dumbbell shape" on coronal & sagittal sequences
Meningioma	Hyperostosis & bone thickening, jugular tubercle	Intense & homogeneous enhancement, dural tail
Fibrous dysplasia	Pagetoid, sclerotic, & cystic forms	Intense enhancement

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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: da Silva. Drafting the article: Vidaletti. Critically revising the article: all authors. Reviewed submitted version of manuscript: Vidaletti. Approved the final version of the manuscript on behalf of all authors: da Silva. Administrative/technical/material support: da Silva, Thibes. Study supervision: da Silva. Video audio editing: Vidaletti.

Supplemental Information

Videos

Video 1. https://vimeo.com/991592386.

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