

## An Extracranial Meningioma Leading to Superior Vena Cava Syndrome

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### Abstract

We present a case of extracranial extension of a WHO grade I meningioma into the right jugular foramen down to the superior vena cava and leading to impaired right atrial venous return. We review relevant literature. This is the only known case of extracranial extension of an intracranial meningioma with benign pathology.

**Keywords:** Meningioma; Extracranial; Metastasis; Superior vena cava syndrome

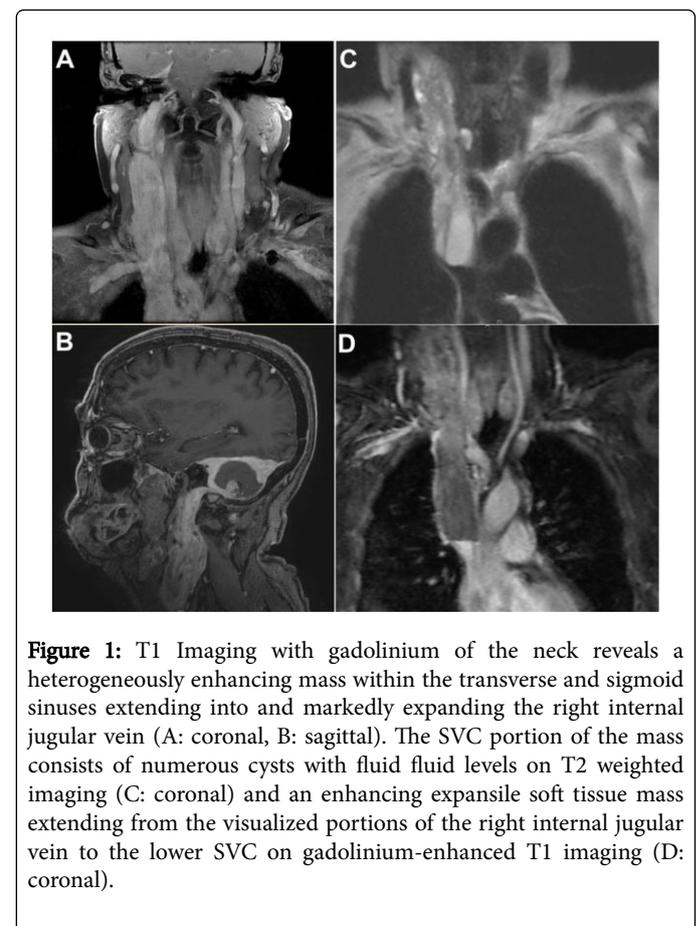
### Introduction

Meningiomas are the second most common primary central nervous system tumors, secondly only to glial tumors. They comprise approximately 25% of all spinal tumors and 15% of all intracranial tumors [1]. Meningiomas only rarely extend or metastasize extracranially, with extracranial presentation comprising only 2% of cases. Intracranial meningiomas are more common in women, whereas extracranial meningiomas are more common in men [2]. There are no distinguishing histologic features between intracranial and extracranial tumors. Due to the rarity of extracranial meningiomas and the lack of localized and specific symptoms, diagnosis of extracranial meningiomas depends on pathological confirmation. We present a rare case of extracranial meningioma extending contiguously from the intracranial space via the internal jugular vein into the right Superior Vena Cava (SVC). This 63 year old woman presented with a painless right neck mass leading to SVC syndrome from obstructed right atrial venous return [3].

### Case Report

We present a 63 year-old, post-menopausal women with extracranial extension of a meningioma presenting with a superior vena cava syndrome by 2014. She initially presented with headaches to an outside facility in 2010, when a brain MRI showed a right posterior middle cranial fossa extra-axial mass with homogeneous enhancement and mass effect on the right temporal lobe and right cerebellar hemisphere. There was internal enhancement of the transverse and sigmoid sinus concerning for sigmoid sinus involvement on initial imaging. She underwent an incomplete resection of a WHO grade I meningioma, pathology was notable for 1 mitotic figure per 10 high powered fields. Ki-67 labelling index of 6% though this included labelling of some mononuclear inflammatory cells. She received involved field radiation for residual tumor in 2011. She was subsequently lost to follow-up. In October 2014, she presented to our facility with severe facial and right arm swelling progressing over several months. Clinical exam revealed facial swelling more on the

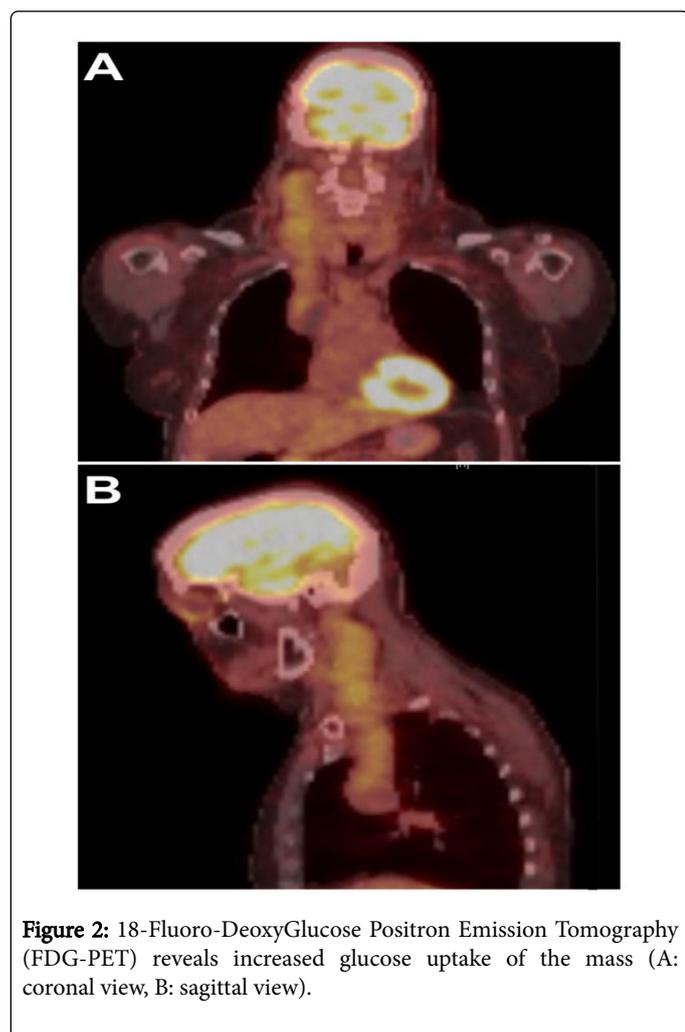
right side, ruddy complexion, periorbital fullness, clubbing of bilateral fingers and engorged chest veins.



**Figure 1:** T1 Imaging with gadolinium of the neck reveals a heterogeneously enhancing mass within the transverse and sigmoid sinuses extending into and markedly expanding the right internal jugular vein (A: coronal, B: sagittal). The SVC portion of the mass consists of numerous cysts with fluid levels on T2 weighted imaging (C: coronal) and an enhancing expansile soft tissue mass extending from the visualized portions of the right internal jugular vein to the lower SVC on gadolinium-enhanced T1 imaging (D: coronal).

A neck and chest MRI and MR Venogram (MRV) revealed an enhancing soft tissue expanding the right transverse sinus extending through the sigmoid sinus and right internal jugular vein, terminating in the Superior Vena Cava (SVC). In the mid SVC, there was a heterogeneous lesion with the appearance of multiple cysts demonstrating T1 hyperintensity and a T2 dependently oriented signal

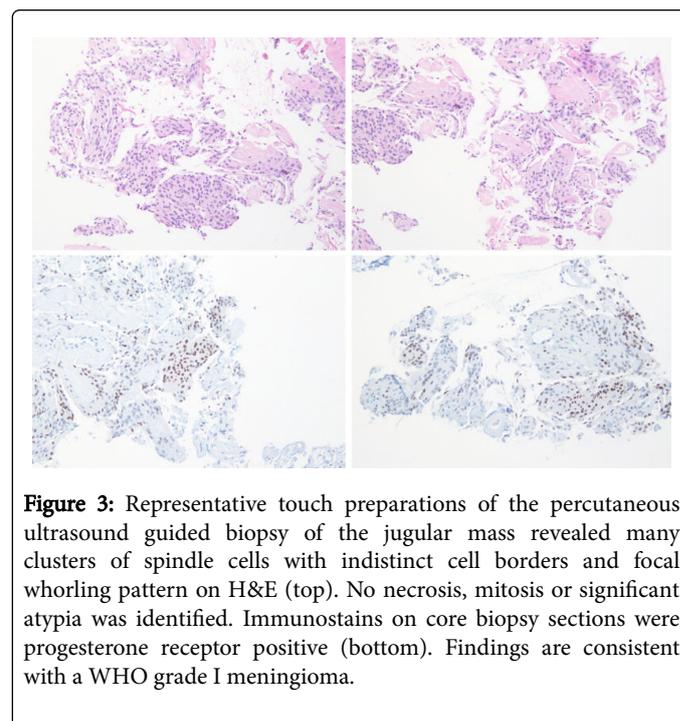
gradient indicating the presence of proteinaceous material or blood products consistent with blood clot. Ultrasound of the neck revealed an expanded right Internal Jugular Vein (IJV) containing a hypoechoic occlusive thrombus with vascularity within the thrombus. A brain MRI revealed nodular enhancement within the right hypoglossal canal and tributaries of the right internal jugular vein likely representing secondary invasion. A calvarial defect in the right temporal bone was consistent with prior craniotomy changes. A head and neck MRV revealed absent flow related enhancement of the right transverse and sigmoid sinuses and internal jugular vein compatible with tumor invasion. A body PET scan revealed mild FDG avidity of the mass. Imaging findings are depicted in Figures 1 and 2.



**Figure 2:** 18-Fluoro-DeoxyGlucose Positron Emission Tomography (FDG-PET) reveals increased glucose uptake of the mass (A: coronal view, B: sagittal view).

A percutaneous ultrasound guided biopsy of the jugular mass was performed. Touch preparations revealed many clusters of spindle cells with indistinct cell borders and focal whorling pattern (Figure 3). No necrosis, mitosis or significant atypia was identified. Immunostains on core biopsy sections were progesterone receptor positive. EMA, CK, CK7, CK20, CK5, p63, TTF-1, S-100, Desmin, CD31, CD34 were negative in the neoplastic cells, supporting the diagnosis of an extracranial meningioma extending to the SVC through the jugular foramen. Second generation genetic analysis revealed three genetic variants: DICER1 c.2614G >A, p.A872T; TERT c.1936C >T, p.R646C;

PDGFRA Variant: NM\_006206.4, c.3155C >T, p.T1052M. DICER1 is a double strand RNA endoribonuclease that functions in gene silencing. In the literature, DICER1 mutations have been associated to rare colon and endometrial tumors, but have not previously been described in meningiomas. TERT encodes telomerase that is responsible for the addition of telomeric repeats. TERT promoter mutations are described in many tumors including aggressive meningiomas and gliomas. PDGFRA is a tyrosine kinase receptor which functions as an important surface signaling factor and mitogen for cells of mesenchymal origin.



**Figure 3:** Representative touch preparations of the percutaneous ultrasound guided biopsy of the jugular mass revealed many clusters of spindle cells with indistinct cell borders and focal whorling pattern on H&E (top). No necrosis, mitosis or significant atypia was identified. Immunostains on core biopsy sections were progesterone receptor positive (bottom). Findings are consistent with a WHO grade I meningioma.

In May 2015, multidisciplinary discussions between neuro-oncology, cardiothoracic surgery, otolaryngology and neurosurgery determined the lesion to be unresectable. In June 2015, she received stereotactic radiosurgery to a symptomatic intracranial portion with prior interval growth leading to progressive headaches, right hemiparesis and right-sided tongue weakness and deviation. She received bevacizumab from June through August 2015. Brain to chest MRI before and while on bevacizumab therapy revealed no further growth of both intracranial and extracranial portions of the meningioma. Her symptoms of right arm and face swelling from SVC compression as well as headaches and right tongue and body weakness improved on bevacizumab. Her treatment was interrupted due to vaginal bleeding related to an intrauterine polyp.

## Discussion

Meningiomas account for 13%–26% of all intracranial tumors, the most common primary brain tumors second to gliomas [4]. As per World Health Organization (WHO) classification, meningiomas are graded as I (benign), II (atypical) or III (malignant) and comprise 80.6%, 15.1% and 4.3% of meningiomas, respectively [5]. Symptoms and complications due to meningiomas are based on their location and mass effect, including seizures, hearing loss, proptosis, cranial nerve deficits and dural venous sinus invasion/ thrombosis. Meningiomas generally tend to invade dural venous sinuses. Extracranial presentation of meningiomas is uncommon, comprising less than 2%

of cases and due to direct extension or metastases typically restricted to malignant WHO grade II tumors [3]. Extracranial meningioma extension can mimic the presentations of schwannomas or glomus jugulare tumors [6,7]. Though previous case reports have reported jugular involvement, our case demonstrates an atypical complication of an intracranial meningioma, extending via the jugular vein into the Superior Vena Cava (SVC) resulting in an SVC syndrome, though with imaging and pathologic findings classic for meningioma.

Extracranial meningiomas can present with neurological dysfunction and cranial nerve deficits as well as sinusitis, proptosis, epistaxis. In some cases, the tumor may present as a neck mass, a bulge in the oral cavity, or a preauricular mass [8]. Meningiomas typically present as dural-based lesion with hyperostosis, isodense-to-hyperdense compared to normal brain and with intratumoral calcification on CT. In extracranial presentations, imaging can be nonspecific though can help narrow the diagnosis. They generally enhance homogeneously and avidly, except for malignant or cystic variants. On MRI, T1 weighted images reveal an isointense/hypointense mass. On the T2 weighted sequence they are isointense-to-hyperintense. Gadolinium-enhanced T1 weighted images reveal intense and homogenous enhancement. Diffusion weighted images depict meningiomas readily due to their increased cellularity. MRV depicts invasion or patency of adjacent dural sinuses prior to surgery. The tumor may infiltrate the dura and adjacent venous sinuses, but it is far less likely to extend into the extracranial draining veins. Often histopathology is required to confirm the diagnosis of meningioma and help direct treatment options [1,2]. Meningiomas involving the jugular foramen are uncommon and pose surgical challenges that may require a staged approach to surgical management [7,8].

There are four general groups of extracranial meningiomas: primary intracranial meningiomas extending directly from the skull, extracranial meningiomas arising from arachnoid cell rests of cranial nerve sheaths, meningiomas having no demonstrable connection with foramina or cranial nerves, and metastases of malignant intracranial meningiomas [3]. This case follows the pattern of the first group. A worse prognosis for extracranial tumors has been associated with women older than 40 years at diagnosis and patients with recurrence or necrosis [8]. In addition, meningiomas of the neck have a propensity towards extracranial expansion, higher recurrence rates due to multicentric growth and occur in combination with other intracranial tumors [9]. Typically, the appropriate treatment for these

patients is complete excision of the meningioma, including bone and soft tissue involvement [10].

In this case, limited surgical options are available given the extent of tumor growth into critical vascular structures. Despite any pathological evidence of malignancy, the rapid, extensive, and extracranial recurrence suggests a poor prognosis despite the WHO grade I histologic classification. We pursued medical and radiation therapy with stable-to-decreased tumor size on MRI and significant symptomatic improvement.

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