The Canadian Journal of Neurological Sciences (2023), **50**, 123–126
doi:10.1017/cjn.2021.239

Canadian Journal of Neurological Sciences
Journal Canadien des Sciences Neurologiques

Practice Pearls

Sporadic Nerve Root Hemangioblastoma: A Rare Neoplasm. Treatment Strategies

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Keywords: Hemangioblastoma; spine tumour; nerve sheath tumour

(Received 1 August 2021; final revisions submitted 7 October 2021; date of acceptance 12 October 2021; First Published online 20 October 2021)

Background

Hemangioblastomas occur both sporadically and in association with von Hippel-Lindau disease and are most common in the cerebellum and spinal cord; however, these tumors can occur anywhere within the nervous system. A rare but described location is along proximal cervical roots, which can present both diagnostic and therapeutic challenges.[1] We describe the multidisciplinary management of a particularly vascular hemangioblastoma receiving its blood supply directly from the V2 segment of the vertebral artery. Surgery without preoperative endovascular treatment could have resulted in significant morbidity for the patient.

Case Presentation

This 57 years old female patient, previously healthy, had a 2-year history of neck and occipital pain. She eventually presented to the emergency department with a particularly severe episode, describing daily, sharp lancinating pain originating in the upper posterolateral aspect of her neck on the left, radiating up over the occiput and down into the shoulder. She was investigated for cardiac ischemia and carotid dissection. Instead, a lesion was identified at the level of the C2 vertebral body on the Computed Tomography angiogram (CTA). In the neurosurgery clinic, the patient described paresthesias in the left hand and subjective weakness. Her left neck and shoulder pain were in keeping with the often vague C3 dermatome. On examination, there was tenderness and spasm in the paraspinal muscles on the left. There was no objective sensory deficit nor weakness but mild slowed fine motor movements in the left hand and a positive Hoffman's sign. Gait was normal but the patient did describe subjectively worsening balance over the previous 2 years

Imaging

The initial CTA demonstrated a vascular lesion abutting the vertebral artery and remodeling both the neural and transverse foramina. Complete MR imaging of the neuraxis confirmed a highly vascular solitary lesion with both an intra and extradural component at the C2–C3 level on the left side (Figure 1A–D). The conventional angiogram confirmed the highly vascular lesion with rapid A–V shunting fed by multiple tiny and short branches "en passage" arising at a 90-degree angle from the V2 segment of the left vertebral artery (Figure 2A).

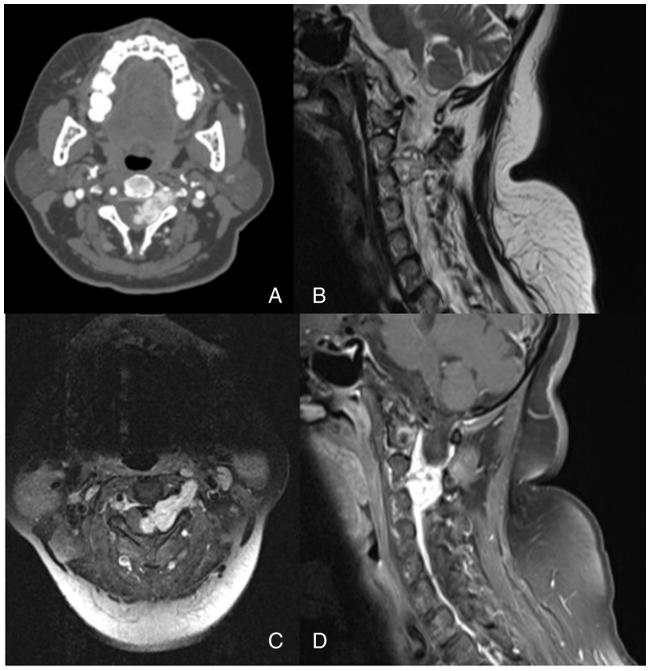

Figure 1: (A) Contrast-enhanced CT demonstrating a lesion in the left neural foramen at the C2–C3 level; (B) the tumor has a heterogeneous signal in T2 and (C–D) avidly enhances following Gadolinium injection and is responsible for significant compression of the spinal cord.

In view of the morphology, location and high vascularity of the mass few differential diagnoses were considered, including nerve root hemangioma, hemangiopericytoma, and less likely

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Cite this article: Tampieri D, Rossiter J, Menard A, and Alkins R. (2023) Sporadic Nerve Root Hemangioblastoma: A Rare Neoplasm. Treatment Strategies. *The Canadian Journal of Neurological Sciences* **50**: 123–126, https://doi.org/10.1017/cjn.2021.239

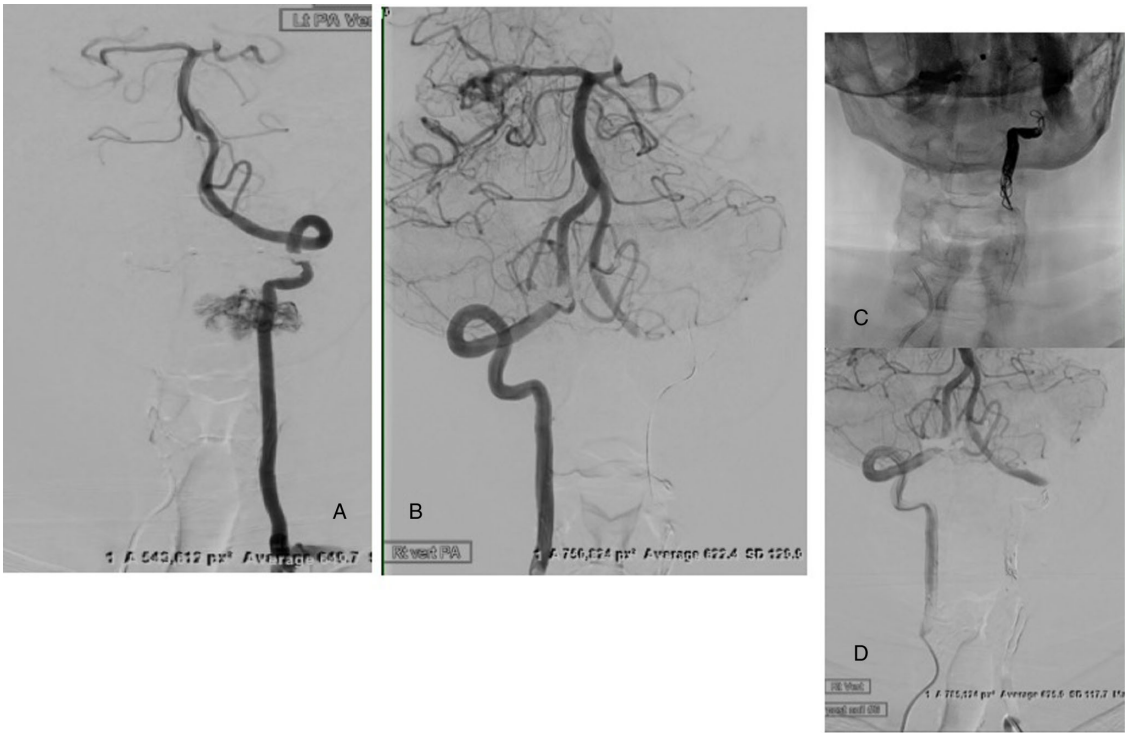

Figure 2: (A) Angiogram of the left vertebral artery demonstrating the hypervascular mass. During the TBO (B), the V4 segment of the left vertebral and left PICA are revascularized in a retrograde fashion. (C–D) Status following left vertebral occlusion using trapping technique with coils.

hypervascular schwannoma. The possibility of hypervascular metastases was not considered, since the patient had no history or primary tumor and there was no destruction of the adjacent bony structures.

Treatment

In view of the highly vascularity of the lesion, as documented by both CTA and MRI, we decided to proceed to conventional cerebral angiography followed by test balloon occlusion (TBO) of the left vertebral. The patient tolerated the TBO of the vertebral artery well, and the angiogram of the right vertebral demonstrated contrast reflux in the V4 and V3 segment of the left vertebral artery with adequate PICA opacification (Figure 2B). Therefore, we proceeded to permanent occlusion of the V2 segment of the left vertebral artery at the level of the tumor to ensure occlusion of the artery and of the vascular supply to the mass (Figure 2C–D).

Following the procedure, the patient was started on ASA. One week later, she underwent posterior instrumented fusion from C2 to C4 with gross total resection of the lesion. Intraoperatively, the lesion was indistinguishable from the C3 root, which was sacrificed intradurally. The tumor had expanded the dural sleeve, extending intradurally where it compressed the spinal cord. Fortunately, there was a preserved arachnoid plane, and only intradural venous drainage without anterior spinal artery supply. Postoperatively, clopidogrel was added for 6 months to mitigate thromboembolic risks. The posterior surgical approach was favored since it enabled a better visualization of the interface between the intradural component of the tumor and the spinal cord. In addition, the significant bone erosion of the lateral aspect of C3 would have resulted in higher risks of long-term instability.

There were no ischemic or other complications. The pathology was that of a hemangioblastoma, WHO grade I (Figure 3A–C). The follow-up MRI demonstrated total gross surgical excision of the mass (Figure 4A–C).

Discussion

Hemangioblastomas are uncommon vascular tumors that can result in significant intraoperative blood loss, with corresponding morbidity. In cystic hemangioblastomas, frequently encountered in the cerebellum, the nodule may be devascularized from within the cyst and resected en bloc. In large and solid hemangioblastomas, circumferential devascularization may be technically very challenging. In these cases, preoperative embolization becomes an important consideration during surgical planning. Controversy exists regarding the routine use of preoperative embolization in intracranial hemangioblastomas due to the elevated morbidity,[2] but if necessary the procedure can be performed in cases of large solid hemangioblastomas.[3] Each case should be discussed by the treating team to identify the best strategy.

We present a case of spinal nerve root hemangioblastoma, and these tumors are rare but we should consider them in the differential diagnosis whenever we encounter high vascular lesions outside the neuroaxis.[1] A systemic review of the English literature of nerve root hemangioblastomas documented a total of 38 case reports, the great majority of the lesions located at the thoracic level; direct surgical excision was the solely modality of treatment.[4] In a recent case report of a cervical extradural nerve root hemangioblastoma, the use of preoperative embolization using particles was described without specific reference to the angioarchitecture of the lesion in relationship with the vertebral artery.[5]

https://doi.org/10.1017/cjn.2021.239 Published online by Cambridge University Press

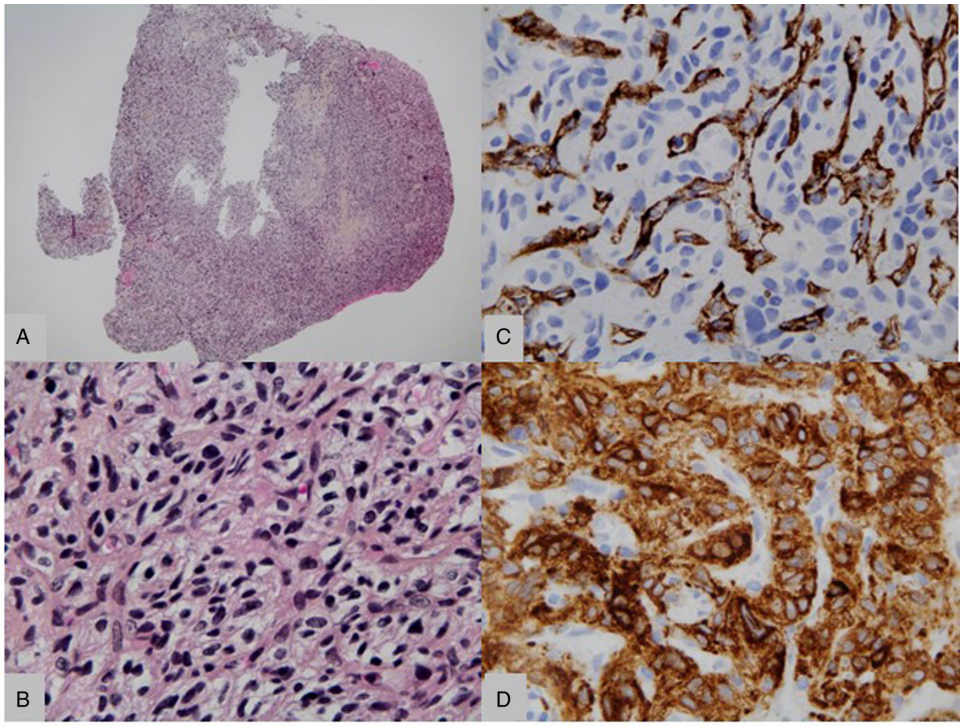

Figure 3: (A–D): Photomicrographs of hemangioblastoma at low and medium power (A, B, HPS stain) showing stromal cells with finely vacuolated cytoplasm, within a dense capillary network that is highlighted by CD31 immunolabeling (C). The stromal cells are strongly immunoreactive for inhibin alpha subunit (D). Scale bar = 500 μm for A, 60 μm for B–D.

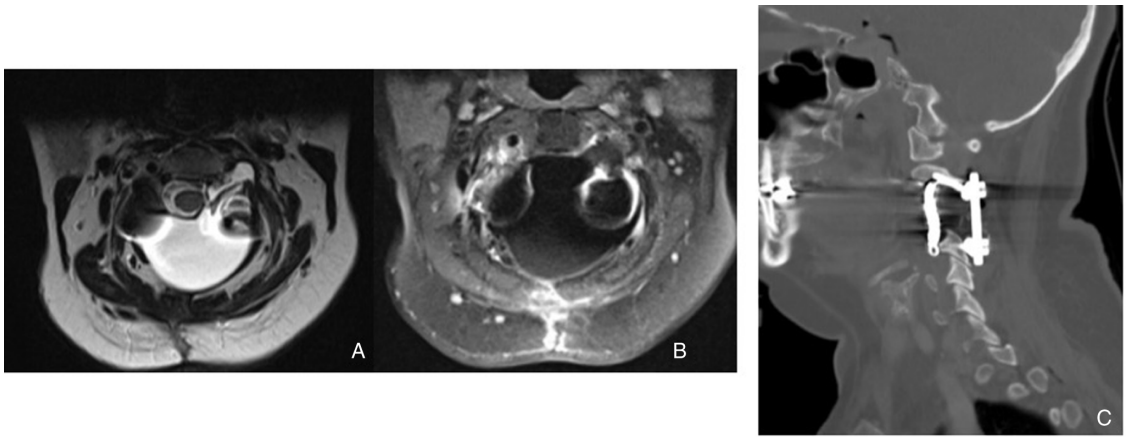

Figure 4: (A–C): Postoperative MRI and CT confirming the total resection of the tumor, the resolved compression on the cord and the surgical instrumentation.

In the case we present, preoperatory embolization with particle or liquid material was deemed not safe due to the tiny caliber of the short segment arteries arising from the V2 segment of the left vertebral artery and supplying the tumor. We felt that an adequate stable position to place the microcatheter in the small arteries supplying the tumor could not be achieved. Additionally, the extra-spinal component of the hemangioblastoma was encasing the left vertebral artery, representing a significant concern during surgical excision. Therefore, we elected to sacrifice the left vertebral artery, following balloon test occlusion, using trapping technique with coils, hence achieving the double goals to devascularize the tumor and occlude the vertebral artery enabling the complete surgical excision.

Disclosures. All authors have no disclosures to declare.

Statement of Authorship. Dr D. Tampieri: primary clinical involvement in endovascular procedure, writing the manuscript, and submitting the paper.

Dr R. Alkins: primary clinical involvement in surgical procedure, and writing and correcting manuscript.

Dr J. Rossiter: pathological diagnosis, discussion and review of pathology, and correcting manuscript.

Dr A. Menard: assisting endovascular procedure.

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