Paraganglioma of the Cauda Equina Presents with Acute Spinal Subarachnoid Hemorrhage and Profound Motor Weakness

Walsh M1, Gordhan A2*, and Emelio Nardone3

1Department of Neurosciences, Advocate BroMenn Medical Center, USA
2Department of Neurosciences, St. Joseph Medical Center, USA
3Department of Neurosurgery, Central Illinois Neuro Health Sciences, USA

Abstract

Paraganglioma of the cauda equina presenting with subarachnoid hemorrhage, lower extremity motor weakness and urinary retention has not been previously reported. Paragangliomas of the cauda equina are confirmed after surgical resection, with no specific imaging criteria for the diagnosis. Imaging characteristics identified in this case report may allow for the tumor to be considered in the differential diagnosis as it may alert the surgeon against excessive manipulation of the lesion, which could lead to hypertensive crisis secondary to catecholamine release. Fluid-fluid levels within the dependent caudal sac by MRI consistent with subarachnoid hemorrhage and the presence of serpiginous vessels on myelographic studies should narrow the differential to paraganglioma versus hemangioblastoma. In addition, CT which has rarely been of use in diagnosing these lesions may be helpful when the presence of hemorrhage is detected.

ABBREVIATIONS

CEP: Cauda Equina Paragangliomas; SAH: Subarachnoid Hemorrhage; NSE: Neuron Specific Enolase; GFAP: Glial Fibrillary Acidic Protein

INTRODUCTION

There are no imaging characteristics that allow for a definitive pre-surgical diagnosis of a spinal cord paraganglioma. We present a case of a hemorrhagic tumor with high peri-tumoral vascularity in the region of the conus and intraspinal subarachnoid hemorrhage (SAH) that was histologically confirmed to be a paraganglioma. A tentative pre surgical imaging diagnosis is important preventing hypertensive crises during resection. Paraganglioma of the cauda equina with the imaging features described, presenting with subarachnoid hemorrhage, lower extremity motor weakness and urinary retention has not been previously reported.

CASE PRESENTATION

A 47 year male presented to the emergency department with abrupt onset of left lower extremity weakness and urinary retention for a period of three days. He denied any other focal neurologic deficit. Neurological exam revealed an alert and oriented patient with normal speech and comprehension. The motor exam identified diffuse weakness in the left leg at 1 out of 5 (flickers or traces contraction upon motor testing). The remainder of his motor exam was intact. The left patellar reflex was absent. Sensation was intact throughout, with no saddle anesthesia. Perirectal sensation appeared intact.

The patient's vital signs were stable. Laboratory studies were unremarkable. CT of the lumbar spine identified a non-specific intraspinal focus of hyper-attenuation at the L1-2 level (Figure 1A, B and C.). MRI showed an ill-defined heterogeneously enhancing intra-dural mass with intermediate T2 signal inferior to the conus. The lesion measured 39 mm in cephalocaudal extent with tumor preponderance within the right para-central spinal canal. Additionally, at the level of S2 an intra-dural fluid-fluid level was identified with intermediate T2 signal inferior to the conus. The lesion measured 39 mm in cephalocaudal extent with tumor preponderance within the right para-central spinal canal. Additionally, at the level of S2 an intra-dural fluid-fluid level was identified with intermediate T1 and T2 signal, consistent with subarachnoid hemorrhage (Figure 2A, B, C, D and E). MR coronal 3-D myelographic source imaging demonstrates an associated prominence of dorsal vascularity cephalad to the lesion (Figure 2F).
A T12-L2 laminectomy for microsurgical resection of the intra-dural tumor was performed. Upon opening the arachnoid layer a dark red sausage shaped tumor was identified and resected completely. Post procedural follow-up MRI showed no evidence residual or recurrent tumor. The patient’s neurologic deficits resolved completely.

The gross pathology of the specimen was described as an elongated dark red tissue sample 3.5 x 1.3 x 1.0 with a smooth surface (Figure 3A). Microscopic evaluation showed a thinly encapsulated lesion, round to ovoid shaped cells demonstrating uniform nuclei accompanied by moderately abundant amount of cytoplasm. The cells were arranged in nests with a trabecular-like configuration (Figure 3B). Regions of infarct-like necrosis were present. The immunohistological staining was positive for synaptophysin and NeuN (neuronal nuclei, a neuroendocrine marker), consistent with a paraganglioma.

**DISCUSSION**

Paragangliomas are neuroendocrine tumors that arise from neuroepithelial cell groups called paraganglia. Adrenal paragangliomas are known as pheochromocytomas and are the most common type accounting for approximately 90% of these tumors. Extra-adrenal paragangliomas can occur anywhere in the body, with 90% located in the head and neck region, specifically the carotid body and jugular bulb, known as glomusjugulare tumors. Rarely do paragangliomas develop in the spine, but those that do tend to be located in the cauda equina and filumterminale. Due to their rarity, cauda equina paragangliomas (CEP) are often omitted from the differential diagnosis of intra-dural tumors in this region, likely due to the fact that CEPs represent approximately 3-4% of lesions in this region [1].

Cauda equina paragangliomas are diagnosed after surgical resection with the assistance of immunohistological staining techniques. Synaptophysin, chromogranin, and neuron specific enolase (NSE) are common stains that are positive in paragangliomas, while glial fibrillary acidic protein (GFAP) staining is negative. Microscopically paragangliomas exhibit the presence of “zellballen,” or a nesting of cell groups, and trabecular cords of cells within the thin fibrovascular stroma. Grossly CEPs present as a soft, dark red and well-circumscribed lesion with attachment to the filumterminale [2].

No definitive MRI imaging characteristics allow for a paraganglioma preoperative diagnosis. MRI is the imaging modality of choice for investigation into spinal tumors with a differential diagnosis of ependymoma (myxopapillary type), schwannoma, hemangioblastoma, meningioma, and metastasis. On T1-weighted images paragangliomas appear

---

**Figure 1** A and B: Non contrast CT of the lumbar spine bone window, sagittal (A) and coronal (B) views demonstrate a punctate hyperdensity (arrows) suggestive of intrathecal hemorrhage. C: Non contrast CT lumbar spine soft tissue window axial section at the level of L1 identifying the punctate hyperdensity consistent with hemorrhage within the right lateral thecal sac (arrow).

**Figure 2** (A, B, C, D, E and F): MRI lumbar spine with and without gadolinium. (A) Sagittal T2 sequence with an extra medullary intra-dural mass at the L1-L2 levels with intermediate increased T2 signal (fine arrow). Fluid-fluid level present at the S2 level is compatible with dependent hemorrhage [thick arrow]. (B) Sagittal T1 sequence with slight hyperintensity related to the mass at the L1-L2 level. (C) Sagittal T1+Gd sequence identifies heterogeneous contrast enhancement of the mass. (D) Axial T2 sequence demonstrating the mass in the right lateral aspect of the cal sac with intermediate T2 signal (arrow). (E) Post contrast axial sequence through the mass showing heterogeneous enhancement (arrow). (F) 3D coronal MR myelogram showing prominent dorsal vascular flow voids at the T12 thought to L2 levels.
hypo- or isointense to the conus medullaris and on T2-weighted images hyperintense signal can be expected. After Gadolinium administration avid enhancement is usual [1-3]. These MRI characteristics are nonspecific because similar imaging features are present for schwannoma and meningioma, making the distinction impossible [1]. The presence of serpiginous flow voids around lesions has been suggested to be associated with highly vascular tumors [4]. Utilizing this vascular association leads to a differential diagnosis that includes hemangioblastoma and vascular neurinoma [5]. In addition to these vascular findings, our patient had a fluid-fluid level in the dependent intra-dural sac with intermediate T1 and T2 signal consistent with subarachnoid hemorrhage. Only one other report described subarachnoid hemorrhage by MR imaging in a pathologically confirmed paraganglioma [6]. Imaging with CT is of no value in diagnosing paragangliomas [7]. Erosion or scalloping of the vertebral bodies or pedicles if seen on plain radiographic imaging or CT is indicative of a slow-growing lesion [6,7]. The intraspinal hyper density consistent with acute blood identified by CT in our patient corresponded with the location of the tumor by MRI. The presence of a tumor related hemorrhagic focus by CT may suggest an imaging diagnosis of a paraganglioma.

Fundamental considerations when operating on paragangliomas is their highly vascular nature as well as their effects on endocrine and cardiovascular function. Post-operative hemorrhage has been identified in one case with devastating results, which has been related to the associated serpiginous vessels of the CEP [8]. It is rare for CEPs to present with clinical manifestations pre-operatively. It is important to recognize that intra-operative tumor manipulation can potentially trigger a hypertensive crisis by excess catecholamine release [3,8].

Imaging characteristics on MRI such as intra-dural dependent fluid-fluid levels suggesting subarachnoid hemorrhage and tumor related serpiginous prominent flow voids imply a highly vascular lesion. With hemorrhagic hyperdensity on CT, a presurgical consideration for paraganglioma can potentially be made. Therefore appropriate preoperative work-up and awareness may assist the surgeon and anesthesiologist in preventing any untoward hemodynamic events during and after surgery.

REFERENCES