Acute Paraplegia From Hemorrhagic Paraganglioma Of Filum Terminale: Case Report And Review Of Literature
T Ma, B Rubin, B Grobelny, D Zagzag, M Koslow, I Mikolaenko, R Elliott

Citation

Abstract
Paraganglioma (PGL) of the filum terminale is a rare tumor of extra-adrenal paraganglia. The reported cases of filum terminale and cauda equina PGLs often present with low-back pain and sciatica. While sensory or motor deficits and paraplegia may occur, the incidence is relatively low. We present a case of a 51-year old male with hemorrhagic paraganglioma of the filum terminale. He presented with acute paraplegia and was treated via emergent laminectomy, evacuation of hematoma, and resection of tumor. The patient had a significant but incomplete neurological recovery. The clinical, radiologic, and histopathological characteristics of the condition are described.

ABBREVIATIONS

INTRODUCTION
Paragangliomas (PGLs) are rare tumors of extra-adrenal paraganglia 2,8,14,15 and occur in the cauda equina or filum terminale in fewer than 4% of cases. 47 The mean age of filum terminale PGL is approximately 40 to 60 years with a slight male predominance of 1.5:1. 6,17,31 The first case of PGL of the filum was described in 1970 and was misdiagnosed as a “secretory ependymoma of the filum terminale” due to a lack of specific histological and immunohistochemical techniques. 29

The most common presentation in lumbar PGL is lower back pain (> 50% of cases) with or without radicular pain into the legs (~20% of cases). 17,28 Sensory or motor deficits, sphincteric and erectile dysfunction and paraplegia may also occur, but their incidence is relatively low. 3,12,17,19,24,33,34,43,49

We present a case of a 51-year old male with hemorrhagic paraganglioma of the filum terminale. He presented with acute paraplegia and was treated via emergent laminectomy, evacuation of hematoma, and resection of tumor. The patient had a significant but incomplete neurological recovery. The clinical, radiologic, and histopathological characteristics of the condition are described.

CASE REPORT
History and Presentation: This 51-year old male initially presented to our clinic complaining of 6 months of progressive lower back pain with radicular pain into both legs down to the feet. He had a history of severe coronary artery disease, underwent coronary artery bypass grafting and was taking aspirin and plavix. On neurologic examination, he had mild weakness in dorsiflexion (4/5) but was without sensory loss or gait difficulty. MRI demonstrated a large intradural tumor extending from L2 to L5 with compression of the cauda equina (Figure 1A) that was well encapsulated, heterogeneously enhancing and had sepentine flow-voids in the subarachnoid space cranial to the tumor. On T2 imaging, the lesion was hyperintense (Figure 1B). On axial MRI, the mass filled the majority of the cross-sectional area of the lumbar spinal canal (Figures 1C & D).
Figure 1
Figure 1. Sagittal T1 (+ gadolinium; A) and T2 (B) MRI images showing a large, intradural mass extending from L1 to L5. Flow voids predominate cranial to the mass indicative of venous congestion or high vascularity of the tumor. Axial T1 (+ gadolinium; C) and T2 (D) images show a large mass that takes up the vast majority of the cross-sectional area of the lumbar spinal canal, obscuring the visibility of the cauda equina. A fibrous stalk is visible attaching the ventral aspect of the tumor to the dorsal aspect of the L3 vertebral body (hypointense on T1 and T2; midline).

CT demonstrated remodeling of the posterior aspects of L3 and L4 vertebral bodies, consistent with a long-standing presence of the tumor. The most common lesions in the differential diagnosis for this intradural, enhancing tumor of the cauda equina included myxopapillary ependymoma, schwannoma and neurofibroma. Metastatic disease was unlikely given the chronicity of his symptoms and bony changes on CT.

Treatment & Postoperative Course: The neurosurgery service recommended surgical excision for relief of symptoms and tissue diagnosis but the patient refused treatment. He returned to emergency room 8 months later complaining of 12 hours of acute lower back pain followed by paresthesias in his lower extremities and progressive weakness. On examination, he had trace movement of his iliopsoas muscles bilaterally but otherwise had no motor function in his legs. He had no sensation to light touch, pinprick or joint position sense below the L1 distribution. He was in urinary retention, lacked sensation to Foley traction, had decreased rectal tone and lacked the anal wink reflex.

Emergent CT imaging demonstrated hyperdense material caudad to the dominant mass, raising the possibility of hemorrhage of the tumor. Moreover, the caudad aspect of the mass behind L5 appears hypodense in relation to the more cranial portion of the tumor, which may suggest infarction or edema.
Figure 2
Figure 2. Unenhanced CT of the lumbar spine shows the hyperdense mass within the thecal sac from L1 to L5 as seen on MRI. However, the caudal aspect of the tumor (behind L5) is hypodense compared to the remainder of the tumor, suggesting edema or infarction and there is hyperdense material caudal to the mass (behind the sacrum), suggesting hemorrhage.

He was given 20 mg of intravenous decadron, transfused platelets to attempt to overcome the effects of antiplatelet agents and taken immediately to the operating room. He underwent L1 to S1 laminectomy and excision of intradural tumor. Prior to dural opening, the thecal sac was noted to be dark blue in color and tense, consistent with underlying hematoma. The dura was initially opening above the tumor to prevent downward herniation of the mass. We noted the egress of bloody CSF under pressure as well as a large, firm, well-encapsulated maroon-colored tumor pushing the dorsal nerve roots out of the dural opening. We extended the midline durotomy caudal and found bloody CSF. Extending the dural opening caudally, we encountered frank subdural hematoma inferior to the mass behind S1.

A dissection plane was carefully developed around the tumor, retracting the nerve roots to their respective sides. The tumor was tethered to the filum terminale, which was identified by the large, tortuous vein running along its length. This was coagulated and cut freeing the tumor from its superior attachment. The tumor was then easily elevated from the intradural space and removed en bloc (Video 1). No episodes of hypertension or cardiovascular changes occurred during surgery. Complete tumor removal was confirmed by postoperative imaging of CT with contrast (Figure 3).
Acute Paraplegia From Hemorrhagic Paraganglioma Of Filum Terminale: Case Report And Review Of Literature

Figure 3
Figure 3. Postoperative CT with contrast showing L1 to S1 laminectomy and complete resection of the tumor.

HISTOLOGICAL EXAMINATION
Histopathological findings were characteristic for paraganglioma (Figures 4 & 5).

Figure 4
Figure 4. Microscopic examination revealed nests of tumor cells, which were separated by a richly vascularized fibrovascular stroma (H&E, original magnification X200).

Figure 5
Figure 5. The tumor was composed of small relatively uniform cells with finely granular basophilic to amphophilic cytoplasm and round to ovoid nuclei with “salt and pepper” appearance of chromatin (H&E, original magnification X400).

The tumor stained positive for chromogranin and synaptophysin immunostains. Sustentacular cells were S-100 protein positive.

POSTOPERATIVE COURSE
By two months from surgery, he has regained full motor power in iliopsoas and quadriceps. He is ambulatory with a walker but has only antigravity power in his distal lower extremities. Sensation has returned in his legs but he still has
Acute Paraplegia From Hemorrhagic Paraganglioma Of Filum Terminale: Case Report And Review Of Literature

no perineal sensation and has not regained bowel or bladder function. One year from surgery, he still has mild weakness in dorsiflexion and uses ankle support orthotics for ambulation. He uses intermittent catherization for persistent urinary retention. MRI obtained at one year following resection shows no evidence of tumor recurrence.

DISCUSSION

PGLs are tumors of extra-adrenal paraganglia. PGLs are sporadic neoplasms, but approximately 1% of cases are autosomal dominant. Nearly 80-90% PGLs occur in the head and neck and typically arise in the carotid body or the glomus jugulare. In the CNS, PGLs may occur in the filum terminale or cauda equina, sella turcica, cavernous sinus, pineal gland, pituitary gland, cerebellopontine angle, and petrous ridge. Paraganglionic cells and the neural crest have a common origin, and during embryogenesis, they migrate along the neural tube. Since paraganglionic cells do not belong within the CNS, it is thought that PGLs result from dysfunction of embryonic paraganglia cell migration or non-regression.

Cauda equina or filum terminale PGL is very rare and only represents 3.8% of cases. Gutenberg and colleagues reviewed 215 reported cases of PGL of the cauda equina and filum. Approximately 60% of cases occurred in males and the mean age at presentation was 44 years (range: 9 to 74 years). The most common symptom was lower back pain (50%), often with radiculopathy (~25%). Others report an even higher incidence of lumbago with or without sciatica. Fewer than 10% of patients had motor or sensory deficits and bowel or bladder incontinence was quite rare (3%). Paraplegia is exceedingly uncommon.

MRI is critical in the diagnosis and treatment of cauda equina or filum terminale PGL. The tumor may appear isointense on T1-weighted images and hyperintense on post-contrast T2-weighted sequences. These highly vascular lesions often have homogenous or heterogeneous enhancement and there is often a serpiginous flow void from vessels associated with the upper pole of the tumor. Additionally, T2-weighted images that reveal a hypointense tumor rim suggest para-magnetic effects from hemosiderin, which signifies prior hemorrhage. Since hemorrhage is uncommon, this low-signal-intensity rim (cap sign) on T2-weighted images is an important finding in the diagnosis of PGL of the cauda equina. Spinal angiography reveals the highly vascularized pedicle and “silk cocoon” appearance that further distinguishes PGLs.

Spinal tumors rarely result in subarachnoid hemorrhage. Li et al. reported the first case of spinal PGL exhibiting subarachnoid hemorrhage shown on MRI. Miliaras et al. concluded that chronic hemorrhage can occur with spinal PGL, and Yang et al. also reported intratumoral hemorrhagic cyst fluid in spinal PGL patients. Intraoperatively, the tumors appear maroon or purple in appearance, are sometimes friable and hemorrhagic, consistent with their highly vascular nature.

Histologically, PGLs exhibit an alveolar pattern and well-formed nests known as “zellenballen.” Sustentacular cells surround fibrovascular stroma, which separate the nests. Immunohistochemistry allows positive stain for neurospecific (gamma) enolase and S100. Chromogranin, synaptophysin, and PGP 9.5 positivity may also occur. However, the absence of GFAP and cytokeratin differentiates PGL from other spinal cord tumors.

Under electron microscopy, PGLs contain secretory granules with amines or peptides.

Complete resection of PGLs often provides durable disease control with a low risk of recurrence. With subtotal resection, 10% of cauda equina PGLs recurred within one year following surgery. Landi et al. advocate long-term follow-up for patients with incomplete resection, and relapse has been reported to occur 30 years from resection. Radiotherapy is often administered to patients with incomplete excision.

To our knowledge, this is the first case of acute hemorrhage of a filum terimale PGL that resulted in paraplegia and its postoperative course. Antiplatelet therapy may have contributed to the spontaneous hemorrhage of this typically vascular tumor. Early removal of suspected intradural PGL should be strongly considered in patients taking antiplatelet or anticoagulation medications. Moreover, in a patient with underlying coronary artery disease, surgical manipulation may cause clinically significant hypertension. Excision of PGLs in patients with limited cardiac reserve may jeopardize patients if they experience dramatic increases in blood pressure and pulse rate intraoperatively. Careful intraoperative monitoring of vital signs and cardiac demand are paramount for maximizing cardiovascular outcomes.

CONCLUSION

We report a case of acute paraplegia from a hemorrhagic
paraganglioma. The patient was treated via emergent laminectomy, evacuation of hematoma and resection of tumor. He had a significant but incomplete neurological recovery. The use of antiplatelet agents in patients with intradural PGLs may increase their risk of hemorrhage and consideration should be given to early resection when symptomatic.

References

1998


Author Information

Tracy Ma, B.A.
Departments of Neurosurgery, New York University Langone Medical Center

Benjamin Rubin, M.D.
Departments of Neurosurgery, New York University Langone Medical Center

Bartosz Grobelny, M.D.
Departments of Neurosurgery, New York University Langone Medical Center

David Zagzag, M.D.
Departments of Neurosurgery, New York University Langone Medical Center

Maxim Koslow, M.D.
Departments of Neurosurgery, New York University Langone Medical Center

Irina Mikolaenko, M.D.
Neuropathology, New York University Langone Medical Center

Robert E. Elliott, M.D.
Neurosurgical Care, LLC