Hemangiopericytoma of the sciatic nerve

Case report

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The authors report the case of a hemangiopericytoma arising in a sciatic nerve. It was found to be invasive within the epineurium but sparing surrounding tissues. Adequate resection required sacrifice of the nerve. Hemangiopericytomas can be added to the short list of mesodermal peripheral-nerve tumors.

KEY WORDS • peripheral nerve • tumor, mesodermal • hemangiopericytoma • sciatic nerve

Most peripheral-nerve tumors encountered in the practice of neurosurgery originate from Schwann cells. These tumors include the benign or malignant schwannoma, the neurofibroma, and the rare granular-cell myoblastoma. Also common are peripheral-nerve tumors of neuroectodermal origin such as ganglioneuromas, neuroblastomas, chemodectomas, and pheochromocytomas. These neoplasms generally arise in the retroperitoneum, posterior mediastinum, neck, and dorsal root ganglia.2 Neoplasms originating from the mesodermal component of peripheral nerves, such as cavernous hemangiomas and angiosarcomas, have also been described but are quite rare.3,4,10 These tumors tend to invade rather than displace a nerve’s fascicles.

We report the case of a hemangiopericytoma arising from within a peripheral nerve. It is, to our knowledge, the first report of such a case. Our single experience sheds light on the resectability of this tumor in the peripheral nervous system.

Case Report

This 27-year-old white female nurse was in her normal state of good health until 9 months prior to admission when she developed the insidious onset of right buttock pain that radiated down the posterior part of her thigh to the lateral aspect of her right foot. She consulted a neurosurgeon who obtained a computerized tomography (CT) scan of the lumbar spine which was reportedly suggestive of a herniated intervertebral disc at L-5. A second opinion was obtained and she was advised to continue with conservative therapy. When bed rest failed to relieve her symptoms, she underwent a right L-5 hemilaminectomy and partial L5–S1 discectomy which also failed to relieve the pain. She then consulted a neurologist who performed electromyography and nerve conduction studies that showed evidence of a severe chronic nerve dysfunction confined to the right lumbosacral plexus. She was referred to Duke University Medical Center for further evaluation.

Examination. Physical examination revealed atrophy of the right leg. The patient had 3/5 strength of the biceps femoris muscle, 1/5 strength of the gastrocnemius muscle, and 0/5 strength of the right tibialis anterior and extensor hallucis longus muscles. There was loss of pinprick and light-touch sensation over the right foot and the posterolateral aspect of the right leg. The right ankle jerk was absent.

A CT scan of the right lower extremity and pelvis showed an enhancing tubular mass originating in the lumbosacral plexus and extending the length of the sciatic nerve distally to the lower third of the femur. Magnetic resonance (MR) imaging also demonstrated a tubular mass along the course of the sciatic nerve (Fig. 1). On the T1-weighted MR image, increased signal involving tissues adjacent to the nerve was suggestive of extension of the mass or, alternatively, edema or blood products.
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Operation. The patient underwent an open biopsy of the sciatic nerve. The enlarged nerve was exposed at the mid-thigh level between the biceps femoris and the semimembranosus muscles. There was no evidence of tumor in the surrounding muscle or soft tissues and the tumor did not appear to violate the nerve sheath. An attempt was made to separate the tumor from the fascicles of the sciatic nerve. Under the operating microscope, groups of fascicles could be separated from the vascular tumor for a short distance but, as the dissection was continued, the tumor was found to completely surround each fascicle (Fig. 2). Microscopic examination of intraoperative specimens showed the tumor to be consistent with hemangiopericytoma (see below).

Given this diagnosis and our inability to establish a plane between the tumor and adjacent nervous tissue, the involved segment of sciatic nerve distal to the sciatic notch was resected along with tumor. Several days later, the patient returned to the operating room for resection of the intrapelvic segment of sciatic nerve involved by tumor. All pathological specimens were returned with the diagnosis of hemangiopericytoma. The proximal and distal margins of the resected nerve were free of tumor.

Postoperative Course. Postoperatively, the patient developed paresthesias in her right foot which were relieved by transcutaneous electrical nerve stimulation. She received physical therapy and at her discharge 1 month after admission she could walk with the aid of a brace.

Pathological Examination. Light microscopic examination of the excised nerve confirmed the intraoperative impression of tumor infiltrating between fascicles of nerve but remaining within the epineurium. The neoplasm consisted of endothelium-lined sinusoidal cavities of variable caliber surrounded by a dense, disorganized array of small cells. The cellular pattern was monotonous with little pleomorphism. Mitotic figures were difficult to find. These characteristics were considered diagnostic of hemangiopericytoma (Fig. 3).

Discussion

The hemangiopericytoma is a rare tumor of mesenchymal origin. This lesion attracted interest in the neuroscience world when Begg and Garret described it in the meninges and considered it to be similar histologically to the “angioblastic meningioma” of Cushing and Eisenhardt. There has been much debate concerning its true cell line, with consensus now generally supporting its independence from meningiomas.

The term “hemangiopericytoma” was first used in 1942 by Stout and Murray to describe a set of “tumors composed of capillary blood vessels with one or more layers of rounded cells (pericytes) arranged about
them." They presented a series of nine patients with these unique vascular tumors in the soft tissues of the head, shoulder, and upper and lower extremities. Later reports of larger groups of patients by Stout and others indicated a predisposition for these tumors to occur in the soft tissues of the thigh and pelvic retroperitoneum, but the tumor has been found throughout the body.

When approaching a peripheral-nerve tumor, a surgeon must know if the tumor infiltrates or simply displaces the majority of the nerve fascicles. A benign schwannoma is a prime example of a tumor which displaces the majority of the fascicles within the nerve. This anatomy allows the tumor to be resected while preserving the continuity of most of the nerve’s fascicles. Conversely, neurofibromas and malignant schwannomas tend to invade a large portion of the nerve’s fascicles and therefore cannot be removed without sacrificing the involved portion of the nerve. Because of the lack of available literature, it was not known whether the tumor harbored by our patient would be invasive or compressive.

Previous cases of hemangiopericytomas appear to have originated from extraneural tissue and to have secondarily involved adjacent nerves. Enzinger and Smith noted in their study of 106 cases that "the majority of the neoplasms were deep seated, either within muscle or intimately attached to muscle, deep fascia, or periosteum." In Stout’s series of 25 cases of hemangiopericytoma, two patients had involvement of the popliteal fossa and one had involvement of the lateral thigh; a "female purebred collie" had tumor in the region of the elbow; however, Stout made no mention of association with nervous tissue. McMaster, et al. in a study of 60 patients, reported only one case of posterior thigh involvement and noted, without mention of intrinsic nerve origin, that the tumor was "intimately associated with sciatic nerve."

In our patient, tumor cells were found infiltrating the endoneurium between the fascicles of the sciatic nerve, yet leaving the epineurium intact. This suggests an origin within the nerve sheath. The invasiveness of this tumor is similar to that of other primary tumors of mesenchymal origin. Hemangiomas arising intrinsically in nerves have been described as invasive, with nerve fascicles "spread over the surface of the mass," and "not resectable without taking a segment of nerve." Rare angiosarcomas of peripheral nerve origin are likewise invasive as two previous reports relate.

To explain the development of this hemangiopericytoma in a peripheral nerve, one might postulate that this tumor arose from pericytes of the vasa nervorum. These are present both within and without the epineurium. Another possibility is the occurrence of aberrant differentiation of normal Schwann cells, as has been described in malignant peripheral-nerve sheath tumors. It is also conceivable that a hemangiopericytoma could have originated in the tissue adjacent to the nerve and then secondarily invaded the nerve, although one would have expected the presence of tumor cells outside the nerve sheath if this were the case.

Hemangiopericytomas are characterized clinically by a high recurrence rate and metastatic potential. Suggested therapeutic modalities have emphasized wide local excisions, including amputation of affected limbs if necessary, and close follow-up evaluation for recurrence. Fortunately for our patient, amputation was not required and we are now following her with studies for reappearance of the tumor.

In summary, the hemangiopericytoma is a rare soft-
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tissue tumor infrequently found in the central nervous system and is reported here as arising from a peripheral nerve. Like a neurofibroma, it was found to be invasive, but only within the epineurium. Full resection required sacrifice of the involved nerve.

References

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