Intramedullary neurilemoma of the cervical spinal cord

Case report

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A case of intramedullary neurilemoma of the cervical spinal cord is reported and a hypothesis for its origin discussed. The importance of pretreatment biopsy is emphasized.

KEY WORDS • neurilemoma • schwannoma • spinal cord • neoplasms

NEURILEMOMA, the specific tumor of Schwann cells, accounts for approximately 30% of primary intraspinal neoplasms. However, its reported occurrence within the substance of the spinal cord is rare; to the authors' knowledge only 10 cases have been previously reported.

We are reporting a patient with this benign tumor in order to emphasize the importance of pretreatment biopsy.

Case Report

This 48-year-old man entered the hospital because of progressive right-sided weakness of 3 months' duration. His past medical history was noncontributory.

Examination. Physical examination revealed marked flaccid weakness of the entire right side, except the face. Muscles of the right shoulder girdle appeared slightly atrophic, and deep tendon reflexes, while not pathological, were more brisk on the right side than on the left. There were no fasciculations. Both plantar responses were flexor. There was some hyperesthesia on the right side, sparing the face, and sensation on the left side was judged normal. Cerebellar and cranial nerve function and mental status were considered normal. Routine laboratory data including analysis of spinal fluid were within normal limits. Plain skull and spine films were negative as were a technetium brain scan and gallium 67 scintiscan of the vertebral column. A cervical Pantopaque myelogram performed via lumbar puncture revealed localized widening of the spinal cord at the C-2 level (Fig. 1 left). In addition, two lobulated filling defects on the right ventrolateral surface of the cord appeared to represent either tumor vessels or dilated spinal veins secondary to local venous compression by the expanded cord (Fig. 1 right).

Bilateral retrograde brachial angiography
was performed, and no abnormalities were noted despite utilization of photographic subtraction techniques.

Treatment and Course. In view of the clinical and radiographic evidence of a high intramedullary tumor of the cervical spinal cord, radiation therapy was begun without benefit of a tissue diagnosis. During therapy the patient developed a probable Brown-Séquard syndrome at the first or second cervical level with right hemiparesis, left hypesthesia, right hyperesthesia, and impaired proprioception on the right side. After receiving 1525 rads to the cervical region during an 18-day period, the patient developed signs of thrombophlebitis of the right leg. Four days later he complained of sudden chest pain, became unresponsive, and died.

Postmortem Examination. An autopsy revealed the immediate cause of death to be a large thromboembolus which obstructed both main pulmonary arteries. The only other significant lesion was in the spinal cord. A firm, well-delimited, solid white nodule, 3 cm in length and 1 cm in diameter, involving the right dorsolateral portion of the first through third cervical segments, was found (Fig. 2). Since no abnormalities of the subarachnoid space were apparent, the extramedullary defects demonstrated on the myelogram were presumed to represent distended veins.

Microscopically the lesion was a...
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moderately cellular neoplasm composed of uniform elongated cells arranged in interlacing bundles and whorls (Fig. 3 left). Focally, nuclei tended to be oriented in palisades (Fig. 3 right). Within the tumor, stout skeins of collagen, scattered clusters of foam cells, and blood vessels with thickened, hyalinized walls were seen. Although sharply circumscribed, the lesion lacked a true collagenous capsule. Continuity of the neoplastic cells with the sheath of the proximal portion of a dorsal nerve root was observed (Fig. 2); the diagnosis was intramedullary neurilemoma.

Discussion

The cellular and extracellular composition, "organoid" pattern, foam cells, and hyalinized blood vessels allowed unequivocal differentiation of this lesion from schwannosis, a non-neoplastic proliferation of Schwann cells within the cord that may be relatively common, and from "pilocytic glioma," a tumor Rubinstein believes is present in most reported cases of "central schwannoma."

Since neurilemoma is a neoplasm of Schwann cells that are not indigenous to the central nervous system, various hypotheses have been advanced to explain the occurrence of intramedullary neurilemomas. These include central displacement of Schwann cells into the spinal cord during embryonic development, Schwann cells ensheathing

Fig. 2. Photograph of transverse section of the spinal cord at the level of the second cervical segment illustrates a well-circumscribed solid tumor in the right dorsolateral portion. A dorsal nerve root is contiguous with the lesion (arrow).

Fig. 3. Photomicrographs of tumor. Left: "Organoid" pattern in moderately cellular neoplasm composed of spindle cells and collagen. H & E, × 68. Right: Palisading of nuclei (center). Note also the abnormal blood vessel with a thick hyalinized wall in the lower left area of the photomicrograph. H & E, × 170.
aberrant intramedullary nerve fibers, and Schwann cells in the sheaths of peripheral nerves accompanying blood vessels into the cord.\textsuperscript{9,7,0,11}

In this case continuity of the neoplastic cells with the sheath of a dorsal nerve root, not apparent by examination of the radiographs or the gross specimen, was observed microscopically. Although this possibly could represent extension from tumor to nerve root, it seems more likely that the neoplasm began at or near the point where a dorsal nerve root, with its sheath containing Schwann cells, pierced the pia-arachnoid and subsequently expanded within the spinal cord. The following evidence supports this hypothesis: 1) most intraspinal, extramedullary neurilemomas are located posteriorly;\textsuperscript{4} 2) most previously reported intramedullary neurilemomas have been located near the periphery of the cord posteriorly;\textsuperscript{4} and 3) nerve plexuses accompanying blood vessels into the cord have been observed exclusively in association with branches of the anterior spinal artery and not with vessels supplying the dorsal aspect of the cord.\textsuperscript{9} If this theory is correct, we might expect to eventually encounter a purely intraspinal "dumbbell" neurilemoma with both intramedullary and extramedullary components. Such a case has not, to our knowledge, been documented.

Despite the paucity of reports, intramedullary neurilemoma may not be as rare as is generally believed.\textsuperscript{8} Since it is a circumscribed, potentially enucleable, and curable neoplasm, its presence should be excluded histologically in the evaluation of all patients with suspected intramedullary spinal cord neoplasms before therapy is instituted.

References

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