Nondural-based lumbar clear cell meningioma

Case report

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This 32-year-old man had noticed right leg pain for 4 years and developed classic right sciatica after heavy lifting, followed by episodes of buckling of both legs 1 month prior to admission. His medical history included congenital left abducens palsy. Examination revealed a right Lasègue’s sign and Fajersztajn’s sign with mild weakness of the right extensor hallucis longus. Magnetic resonance imaging revealed a 1.5 × 2.0-cm enhancing intradural lesion at the L3–4 level. Following laminectomy of L-3 and L-4 and intradural exposure, the tumor was found to be draped loosely by the roots of the cauda equina and attached to a single root without any adherence to dura. Transection of the adherent fascicles and typical microdissection of arachnoidal filaments permitted its complete removal without violation of the capsule, allowing the preservation of a large fascicle. The patient’s recovery was uneventful. Postoperatively, a mild right lateral foot hypalgesia and diminution of the right ankle jerk implicated the S-1 root. Histological and immunohistochemical analyses diagnosed the specimen as a clear cell meningioma.

KEY WORDS • lumbar meningioma • clear cell meningioma • spinal tumor

LUMBAR meningiomas are rare. When they occur, as is the case with thoracic and cervical meningiomas, they are characteristically invested with arachnoid and dural based. We present an unusual instance of lumbar meningioma characterized by absence of dural attachment, dense adherence to the fascicles of the right S-1 nerve root, and a histological and immunohistochemical appearance that led to the diagnosis of a clear cell meningioma.

Case Report

This 32-year-old right-handed man initially experienced right leg pain 4 years previously and thereafter experienced intermittent pain episodes of short duration. One month prior to admission, after heavy lifting, he suffered a low-back sprain and recurrent right sciatica radiating posterolaterally to the dorsum of the foot. Shortly afterward he began to have episodic buckling of both legs. Turning his head to the right provoked the right sciatica, as did a recumbent posture.

His medical history included removal of two dermatofibromata from the skin of his right anterior thorax and right shoulder and a left congenital abducens palsy.

Examination. Examination disclosed left abducens palsy, weakness of the right extensor hallucis longus, and impaired straight leg raising on the right to 10° and on the left to 40° with crossed leg pain. There was mild lumbar tenderness to punch.

Magnetic resonance (MR) imaging demonstrated a poorly delineated, nearly isointense elliptical lesion at L3–4 on T₁-weighted imaging, which became dramatically outlined on T₂-weighted imaging (Fig. 1 left) and clear-
ly enhanced with gadolinium (Fig. 1 right). The appearance was that of an intradural lesion.

**Operation.** Lumbar laminectomy of L3–4 and intradural exposure disclosed a 1.5 × 2.0–cm elliptical lesion adherent to a right-sided nerve root. One fascicle of the root was lightly adherent to the dorsal aspect of the capsule and could be dissected free and preserved. The other fascicles split, traversed both sides of the capsule and then rejoined. There was no dural adherence whatsoever. The lesion’s appearance at operation was that of a typical neurofibroma. The fascicles surrounding the tumor were transected. The tumor was separated from filmy strands of arachnoid by microdissection and then removed intact without violation of its capsule.

**Postoperative Course.** The patient’s postoperative recovery was uneventful. Neurological examination immediately after surgery and at 1 month demonstrated right S-1 dermatomal hypesthesia and diminution of the right ankle jerk without any weakness. Postoperative MR studies of the brain, cervical spine, thoracic spine, and lumbar spine revealed no residual tumor at the operative site and no other tumors.

**Pathological Examination.** Grossly, the tumor consisted of a round, well-circumscribed firm pale-gray nodule measuring 1.8 × 1.3 × 1.3 cm.

Microscopically it was characterized by sheets and vaguely defined nests of cells separated by delicate vascular channels. Hyaline fibrosis was present, varying from minimum centrally to marked peripherally. The cells had abundant clear cytoplasm with round uniform bland-appearing nuclei without mitoses (Fig. 2 upper left). Occasionally the cells formed small whorls, as is characteristically seen in more typical meningiomas (Fig. 2 upper right). Nerve trunks were present within the tumor (Fig. 2 lower left) and compressed large nerve trunks were present surrounding the tumor (Fig. 2 lower right), dura was not seen.

Histochemical staining revealed the presence of a small-to-moderate amount of periodic acid Schiff-positive diastase-sensitive granules consistent with glycogen. Mucicarmine and Alcian blue stains were negative for mucin.

Immunohistochemical examination revealed the tumor to be positive for vimentin and Leu 7 and weakly positive for epithelial membrane antigen. It was negative for cytokeratin, S-100 (the surrounding nerve roots were strongly S-100 positive), glial fibrillary acidic protein, Ham 56, and Mac 387. Immunohistochemical analysis also revealed the tumor to be progesterone receptor positive and estrogen receptor negative.
Electron microscopy revealed the presence of occasional desmosomes and abundant intermediate filaments, and flow cytometry determined that the tumor was aneuploid with a 3% S-phase fraction. These findings are considered to be typical of a clear cell meningioma.2

Discussion

Clear cell meningioma is one of the histological forms of meningioma. Reference to this tumor has been made previously.1,2 In a third publication, Zorludemir, et al.,3 detail the cases and pathology reviewed and analyzed at the Mayo Clinic that were mentioned in one of the previous publications.2

To date, approximately 14 cases have been described with six (43%) spinal intradural tumors, of which five were lumbar and one thoracic.2 One of the cases was “also primarily nerve-associated with no significant or minimal tenuous attachment to dura” (BW Scheithauer, personal communication, 1995). It was stated for the entire series, including four (26%) tumors in the cerebellopontine angle, three (21%) in a supratentorial location, and one (7%) at the foramen magnum, that “despite their benign appearance, they are inordinately aggressive,” with a 50% recurrence rate that includes local recurrence in 21% and spinal metastases in 7%.2

The clinical presentation, neuroimaging features, and especially the operative findings in our case were more typical of a neurofibroma than a meningioma in view of the lack of a dural attachment, making this tumor unlike any spinal meningioma we have previously encountered.

The importance of recognizing this particular variant of meningioma relates specifically to its potentially more aggressive clinical course.2

The absence of other tumors on postoperative MR imaging in our patient mitigates against remote seeding or local dissemination as a cause of the tumor and points to its arachnoidal cell origin.

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References


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