

## Thoracic Intramedullary Clear Cell Meningioma

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Intramedullary clear cell meningioma (CCM), which is more aggressive than other variants of meningioma, is extremely rare. To date, only one case has been documented in spinal tumors. We report the first case of an intramedullary CCM originating in the spinal cord at the thoracic region.

**KEY WORDS :** Clear cell · Meningioma · Intramedullary.

### Introduction

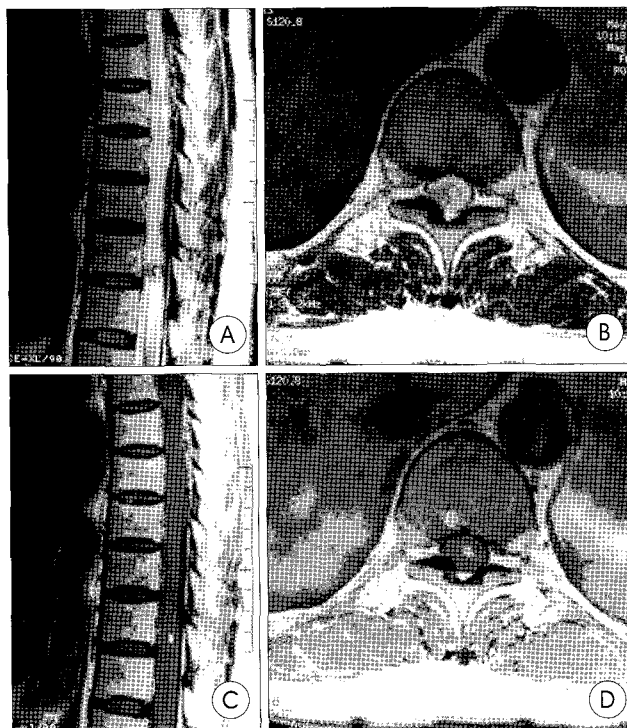
Spinal meningiomas are relatively common spinal intradural tumors and comprise approximately 25% of intraspinal neoplasm<sup>9)</sup>. Of them, however, intraspinal clear cell meningiomas (CCMs) are rare histological variants of meningioma. As far as we know, fewer than 20 cases have been reported to date<sup>3-6,12,15)</sup>. Also, intramedullary spinal tumors are rare and account for only 5% of spinal tumors<sup>1)</sup>. Intramedullary meningiomas of the spinal cord are extremely rare lesions of the spine. In our review of English literature, we could find only six cases that have been reported<sup>2,6,10,11,13)</sup>. Only one of those six cases reporting histological diagnosis was intramedullary CCM. Upon the reviewing the case reports, we found that our own is the first case report of a thoracic intramedullary CCM. We present a case of intramedullary CCM, a rare clinical entity, and review the literature that has been previously reported.

### Case Report

A 65-year-old woman had a 2-month history of progressive leg paresthesia and weakness. She suffered a progressive gait disturbance and a sensory disturbance on and below her T10 dermatomes. Bilaterally, her triceps, knee, and ankle reflexes were accelerated. The neurological findings indicated that her condition was consistent with T10 myelopathy.

Spinal magnetic resonance (MR) imaging revealed that the thoracic spinal cord was diffusely enlarged with an extensive intramedullary spinal cord abnormality extending from the

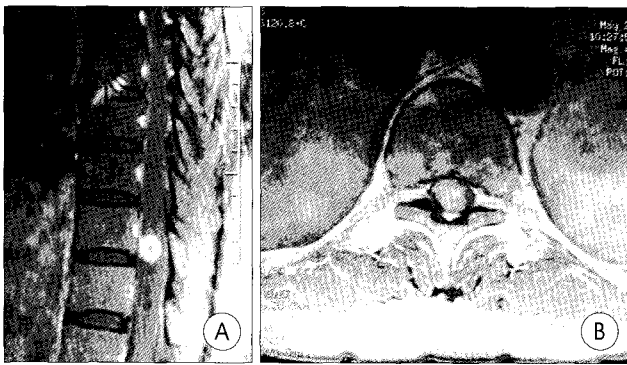
T6 to the T10-11 intervertebral disc space (Fig. 1). The tumor was posterior to the T9-10 vertebral levels and had isointense signals with a high signal central spot on T1- and T2-weighted sequences. There was a mild expansion associated edema situated superior and inferior to the lesion. The tumor enh-



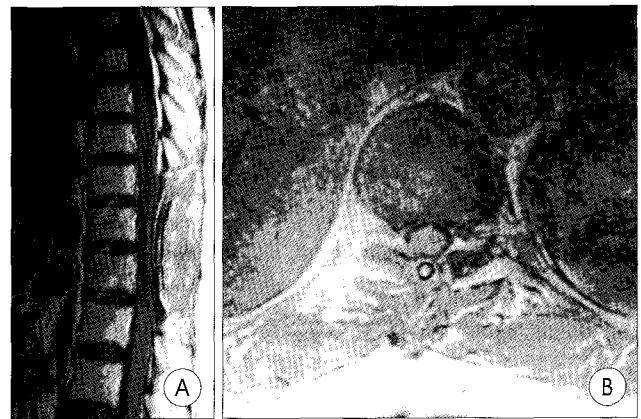
**Fig. 1.** T2- (A, B) and T1- (C, D) weighted magnetic resonance images revealing edematous spinal cord change. At T9-10, an intramedullary, iso signal intensity lesion is detected on MR images.

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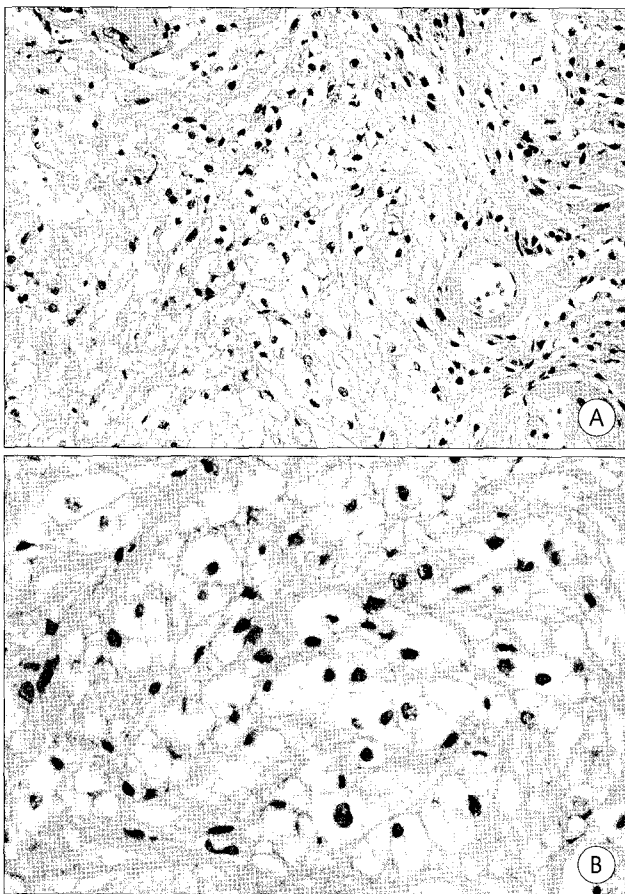
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**Fig. 2.** T1-weighted magnetic resonance images with gadolinium enhancement, showing a homogeneous enhancing intramedullary tumor. Axial gadolinium-enhanced T1-weighted magnetic resonance image obtained at T9-10, demonstrating the enhanced lesion almost at the center of the spinal cord.



**Fig. 4.** Immediate postoperative T1-weighted magnetic resonance sagittal (A) and axial (B) images with gadolinium enhancement showing no residual tumor.



**Fig. 3.** Photomicrographs of the spinal lesion under low magnification (A, B), demonstrating clear cells with round to oval shaped central nuclei and abundant clear cytoplasm, are organized in whorls and interspersed with hyaline sheets (H&E, X20(A), X40(B)).

anced homogeneously with gadolinium infusion (Fig. 2). The finding was consistent with an intramedullary tumor of the spinal cord.

The patient was started on a course of intravenous corticosteroid therapy and subsequently underwent a T9-11 laminectomy for resection of the tumor. The dura mater was opened

and the tumor was intramedullary in location without dural attachment. A myelotomy was performed, and under high magnification a white, hard lesion without cyst or necrosis was encountered and carefully dissected from the surrounding tissue. The lesion was completely resected and the specimen was submitted for pathological examination.

Upon histological examination, the tumor cells revealed round-to oval-shaped central nuclei and abundantly clear cytoplasm with distinct cell borders (Fig. 3). The cells were infiltrated into the cord tissue. The cells showed the immunoreactivity consistent with meningioma cells, such as being epithelial membrane antigen(EMA) positive, vimentin positive, and glial fibrillary acidic protein(GFAP) negative. And they also revealed periodic acid Schiff(PAS) positive and diastase labile PAS(D-PAS) negative features. These findings were consistent with CCM.

The patient's condition rapidly worsened during the immediate postoperative course. She became transiently paraplegic and was presented with anesthesia below L1 dermatome. Immediate postoperative MR imaging of the spine revealed no residual tumor (Fig. 4). Wound revision was completed and a focal swollen spinal cord was found. The wide laminectomy on T7,8,12 and duroplasty were done and neurological deficits improved significantly within the first month. She continued to experience some sensory deficits and weakness in both legs required short stay at the rehabilitation facility.

No adjuvant therapy was administered. One year postoperatively, the lower-extremity strength had recovered to good state except for severe weakness (2+/5) of the left leg.

## Discussion

Spinal meningioma in the intramedullary compartment is extremely rare. Only six cases have been reported in the English literature, all of which ranged in age from 22 months

to 69 years<sup>2,6,10,11,13</sup>). The two cases -the case of Jallo et al.<sup>6</sup> and our own- showed an intramedullary CCM. The CCM variant has only recently been described and studied<sup>15</sup>).

As for the location of spinal meningiomas at the spinal level, cervical meningiomas account for 14 to 18.5%, thoracic for 75 to 82%, and lumbar for 2 to 7% of these tumors in previous report<sup>9</sup>. Spinal meningiomas are usually found in the thoracic spine. But, after reviewing case reports of the intramedullary meningiomas, four were in the cervical, and two, including our own, were located at the thoracic spine. This seems to show a possible tendency of these tumors to present themselves in the cervical spinal cord.

Our case involved the thoracic spinal cord and contrasted with the preferred site of intramedullary meningiomas, which is the cervical region. Thus, the present case is the second report of a thoracic intramedullary meningioma as well as an intramedullary CCM. And this is the first case report of a thoracic intramedullary CCM.

The pathogenesis of intramedullary meningioma remains enigmatic. The hypothesis has been advanced to explain the histopathogenesis of intramedullary meningiomas. Shuangshoti et al.<sup>14</sup> suggested that these tumors might originate in the mesenchymal cells lining the perivascular spaces of the neuraxis.

The rarity of the lesion and the nonspecific imaging characteristics make the diagnosis difficult. The differential diagnosis considered in our case, depending on the location and the frequency, included neoplastic conditions such as astrocytomas, ependymomas, or hemangioblastomas<sup>11</sup>. No typical MRI appearance of these tumors was found in our case. However, it is very difficult to distinguish intramedullary meningioma from the other intramedullary lesions without performing a histological examination. Thus, definite diagnosis is only provided by the pathology.

The majority of spinal meningiomas have been reported to be either meningiothelial (29 to 59%) or psammomatous (21 to 57%)<sup>9</sup>. In our study, there was a higher incidence of CCM in intramedullary meningiomas compared to other spinal meningiomas. Intraspinal CCMs are rare histological forms of meningioma, with fewer than 20 intraspinal cases of CCM reported in the literature<sup>3-6,12,15</sup>. The proportion of spinal to total meningiomas is higher for CCM (45%) than for all meningiomas (0.2%). Ordinary meningioma is likely to occur in the fifth and sixth decades of human life with female predominance, however, in CCM, there is a wide variety of age distribution without gender predilection<sup>7</sup>.

Intraspinal CCMs typically are located in the lumbar region and in the subarachnoid space<sup>3-5,12,15</sup>. In contrast, the case of Jallo et al.<sup>6</sup> was located in the intramedullary cervical spine and our own in the intramedullary thoracic spine.

Similar to the pathology of clear cell meningioma on the

brain, our case had distinctive histologic features consisting of a sheet-like or lobular proliferation of often polygonal cells with clear cytoplasm<sup>7</sup>. The new WHO classification of tumors of the central nervous system divides meningiomas into grades I, II, and III by likelihood of recurrence and grade<sup>8</sup>. Among grade II variants, CCMs have been called glycogen-rich meningioma, and were first described by Manivel et al. to represent 0.2% of all meningiomas<sup>15</sup>. This subtype appears to show more aggressive clinical evolution, which is apparently independent of other conventional histological parameters (e.g., mitotic activity, invasion) that usually indicate a risk of such behavior<sup>3</sup>.

Spinal meningiomas are well-circumscribed and slow-growing tumors that are associated with good patient outcomes following surgery. Advances in imaging and surgical techniques have brought about better surgical and clinical results from spinal meningiomas. Nevertheless, intramedullary meningiomas still bear a poor prognosis and intramedullary CCM may be more aggressive. Surgical excision is the treatment of choice in patients with intramedullary meningiomas and appears to be curative.

Excellent results have been reported after complete or partial removal of intramedullary meningioma<sup>6,10,11,13</sup>. As for intramedullary CCM, a 42% local recurrence rate after gross total resection was observed in previous report<sup>6</sup>. Zorludermir et al. reported that CCM showed a 61% recurrence rate, including 15% local discontinuous spread and 8% widespread cranial to spinal metastasis in their 13-case series<sup>15</sup>. Complete surgical removal is necessary because, despite their benign histological appearance CCM are potentially aggressive and may recur, spread locally, and even metastasize. Radiosurgery and radiotherapy have been generally considered as a treatment option in recurring cases. Total removal was performed on our patient, and unfortunately the tumor's excision led to the patient's neurological deterioration. No further adjuvant chemotherapy or radiotherapy was performed.

Due to the rarity of the intramedullary CCM, before surgical exploration they may be difficult to differentiate from other intramedullary lesions. Since complete surgical removal is not attempted in some cases such as gliomas, correct diagnosis from a biopsy specimen is crucial, and it is therefore important to preoperatively recognize of the possibility of the occurrence the intramedullary CCM.

## Conclusion

The authors report a case of thoracic intramedullary CCM without dural attachment. It deserves a certain interest due to its rarity and can be also considered in the differential diagnosis of intramedullary tumors.

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