Clear Cell Meningioma of Cauda Equina in a 10-year-old Child

〈**Case Report**〉

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ABSTRACT

A case of multiple clear cell meningioma of cauda equina in a 10-year-old girl is reported. She was referred with 3-month history of painful episodes of backache and a markedly sore right leg and admitted for further examination. No neurological deficiency was recognized in physical examination. Magnetic resonance imaging scan clearly exhibited heterogenous tumors at the level of Th12 and L2 following roentgenogram that did not show any abnormal findings. Tumor resection was performed by thoraco-lumbar en block laminectomy with T-Saw. Subsequently, resected laminas were replaced to obviate possible spinal kyphosis. The patient's recovery was uneventful. Clear cell meningioma was discerned by pathological examination, whose findings including granules consistent with glycogen and ultrastructure of meningioma.

Younger patients who undergo multiple laminectomy have been reported to incur kyphotic change at involving the spine. In order to obviate such deformities, we think that the posterior element of the spine should be reconstructed.

Key words : Clear cell meningioma, Spinal tumor, Spinal reconstruction

INTRODUCTION

Clear cell meningioma has recently been identified and included in the World Health Organization (WHO) classification as a peculiar variant that differs from conventional meningioma by affecting younger patients, arising more often in spinal or cerebellopontine locations and showing a higher recurrence rate[1,2]. However, It is obvious that the rarity of this tumor increases the difficulty of correct diagnosis[3].

Lumbar meningioma are less common[4]. When they occur, as is the case with thoracic and cervical meningiomas, they are characteristically invested with arachnoid and dural mater.

The present case report illustrates a rare case characterized by unusual multiple lumbar lesions, absence of dural attachment, dense adherence to the fascicles and a histological and immunohistochemical appearance that led to the diagnosis of clear cell meningioma.

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CASE REPORT

A 10-year-old girl was referred with a 3-month history of painful episodes of backache and a markedly sore right leg. Symptom had progressed and finally led to hospital admission for detailed examinations and possible surgical treatment.

Examination

Physical examination revealed that there was no neurological deficiency, including muscle weakness, sensory disturbance, bowel-bladder disfunction, morbid reflex or tension sign of nerve root.

Roentgenogram did not show any abnormal findings. None of the following features were observed ; abnormal interpedicle distance, pedicle sign on antero-posterior view of lumbar spine nor scalloping of vertebral body or enlargement of the foramen on lateral view (Fig. 1).

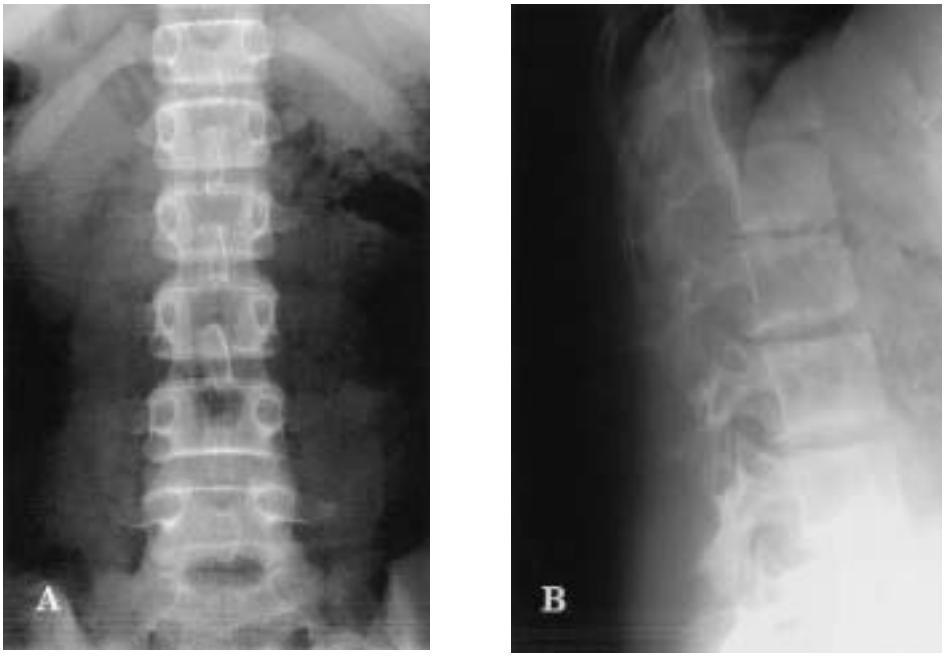

Fig. 1 Roentgenogram did not show any abnormal finding. 1A: antero-posterior view of lumbar spine: 1B: lateral view of lumbar spine.

In magnetic resonance imaging scan, heterogenous tumors of about 2.0 cm in diameter were recognized on the Th12 and L2 levels of spine. Tumors with iso signal on T1 weighted imaging and low signal on T2 weighted imaging with well contrast-enhancing were recognized (Fig. 2).

Myelogram revealed the shadow of a intradural tumor with complete blockage of enhancement of dural sack at the level between L 2 and 3 (Fig. 3).

Surgical Procedure

Thoraco-lumbar en block laminectomy from T12 to L3 was performed with T-Saw[5] with view to later lamina reconstruction. Sequentially intradural exposure disclosed a 1.0 × 1.5 cm elliptical tumor at the level between T12 and L1 and a 2.0 × 3.5 cm elliptical tumor at the level between L2 and L3. The cephalad tumor was strongly adhered to two fascicles and it was impossible to dissect the tumor from nerve tissue. On the other hand, the caudal tumor was lightly adhered to nerve and therefore easy to separate. Both tumors had no dural adherence whatsoever. After transecting the fascicles surrounding the tumors, tumors with involved fascicles were removed by microdissection without violation of the capsule.

Subsequently, resected laminas were re-

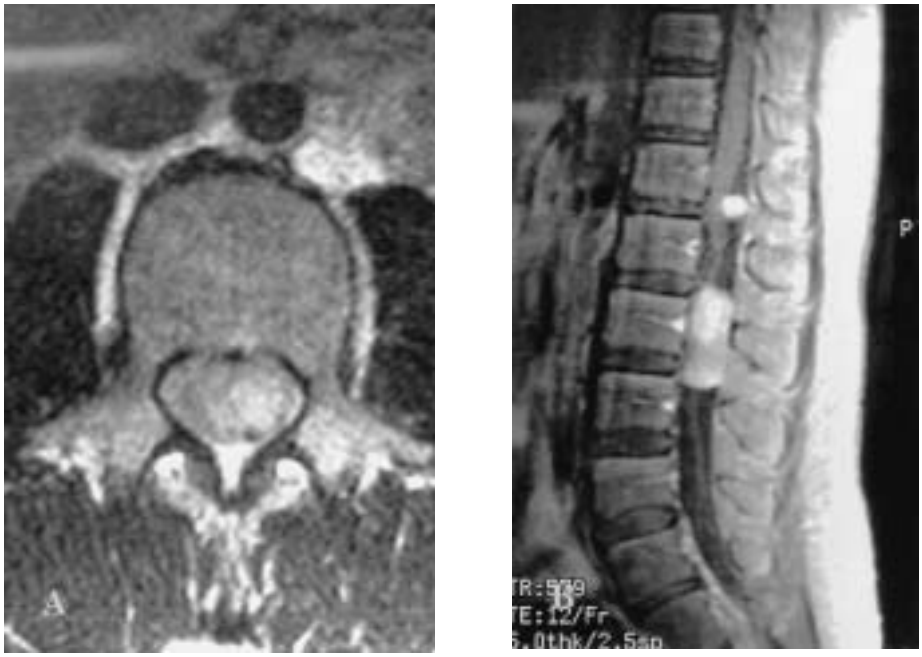

Fig. 2 MRI reveals multiple spinal tumors at the level of Th12 and L2. 2A: tumor at L2 level with low signal on T2 weighted imaging on axial view. 2 B: well enhanced tumors at the level of Th12 and L2 on saggital view.

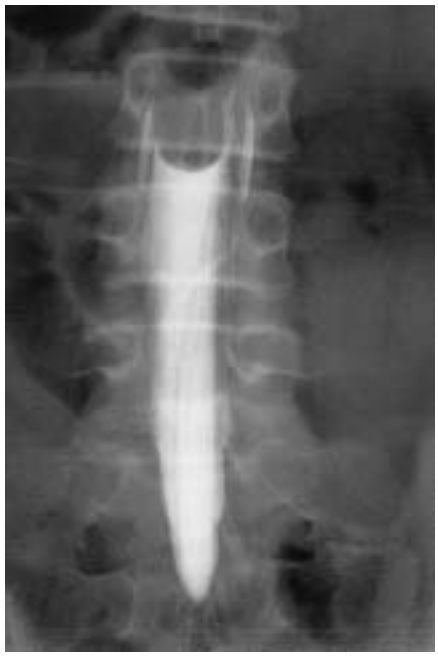

Fig. 3 Myelogram exhibited the shadow of a spinal tumor with complete block of enhancement at L2/3 level.

placed and attached firmly to base bone with nylon suture for the precaution of possible spinal kyphosis (Fig. 4).

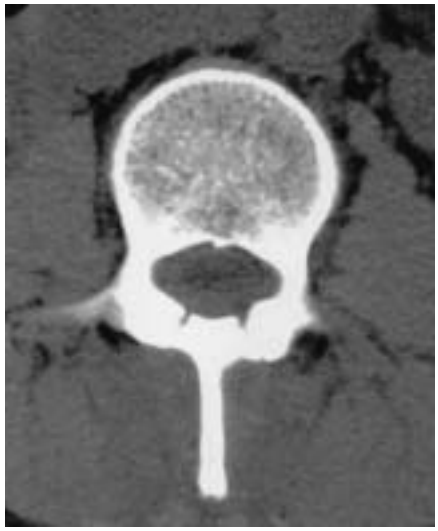

Fig. 4 CT scan shows recapped lamina on L2.

Postoperative Course

The patient's postoperative recovery was uneventful. Postoperative magnetic resonance imaging study of the lumbar spine at 2 years after surgery demonstrated no residual tumor at the operative site and no recurrence.

Pathological Examination

Grossly, the two tumors consisted of elliptical , well-circumscribed firm pale-gray nodules measuring 1.5 × 1.0 cm and 2.0 × 3.5 cm respectively.

Microscopically they were characterized by sheets and vaguely defined nests of cells. Hyaline fibrosis was present. The cells had abundant clear cytoplasm with round uniform bland-appearing nuclei without mitoses. Occasionally the cells formed small whorls, as is characteristically seen in more typical meningiomas (Fig. 5).

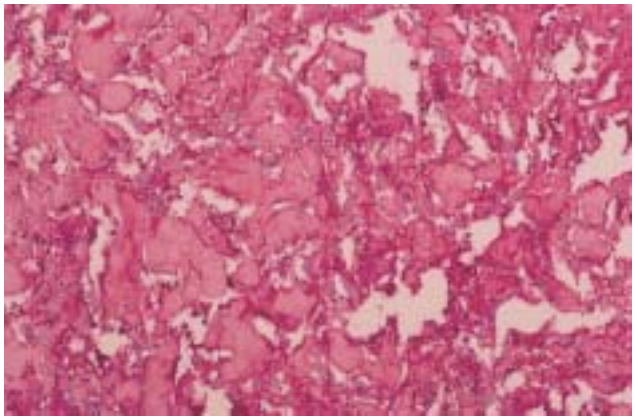

Fig. 5 Light microscopic appearance of tumor (PAS, × 200). The cells had abundant clear cytoplasm with round uniform bland-appearing nuclei.

Histochemical staining revealed the presence of a small-to-moderate amount of periodic acid Schiff-positive diastase-sensitive granules consistent with glycogen (Fig. 6). Immunohistochemical examinations with vimentin, EMA and S-100 were less positive.

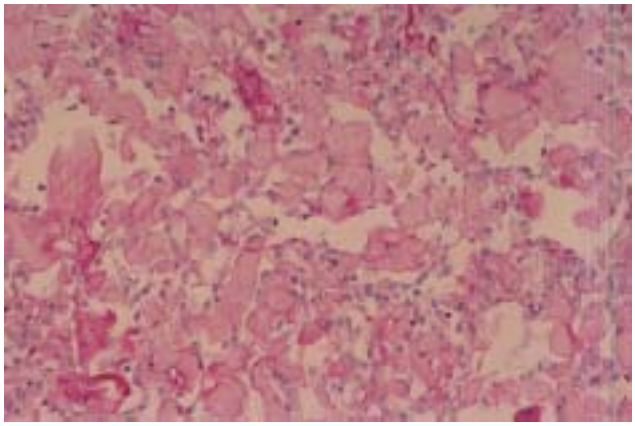

Fig. 6 Histochemical staining revealed the presence of periodic acid Schiff-positive diastase-sensitive granules consistant with glycogen (PAS, × 200).

On ultrastructural examination, the tumors were characterized by moderately abundant cytoplasm literally filled with glycogen particles. Also interdigitation of cell membranes were recognized. The cells were joined by junctional complexes, most often well-formed desmosomes, characterized as meningioma (Fig. 7).

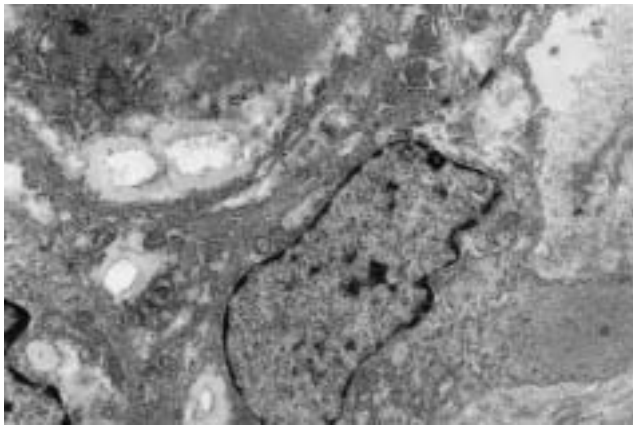

Fig. 7 Ultrastructure of tumor (× 4000). Remarkable cytoplasmic glycogen depositions including variable intermediate filament, imbrication of cell membranes and well-formed desmosomal junctions were recognized.

Based on pathological examinations, the tumors were diagnosed as clear cell meningioma.

DISCUSSION

Clear cell meningioma is an uncommon, aggressive and morphologically unique variety of meningioma that has been reported to occur in the spine and cerebellopontine angle[6,7,8]. It shows no sex predilection and affects young patients more often than conventional varieties of meningioma[7]. It seems to be a more aggressive tumor, with higher incidence of recurrence. Characteristically, this tumor is composed of uniform clear cells and has a prominent hyalinized collagenous stroma[2]. Typical light microscopic and ultrastructural features indicative of meningothelial differentiation are usually sparse or almost completely absent in clear cell meningioma[9].

Clear cell meningioma is an exceedingly

rare tumor of meninges. Only 14 cases have been described, six of which were spinal intradural tumors, five lumbar and one thoracic[3,10,11]. Our case is clearly rare at the point of younger age and multiple involvement of lumbar spine.

Generally speking, under light microscopic observation, there is an obvious lack of meningiomatous features, thus making diagnosis difficult. It is reported that ultrastructurally the tumor have features common to all meningiomas, such as junctional complexes, interdigitation of cells and well-structured desmosomes, which are very helpful to assess the correct diagnosis[7,12]. In our case, the glycogen in cytosol with periodic acid Schiff-positive diastase-sensitive granules and characterization of the ultrastructure in spite of negative reaction in immunohistochemical examinations were helpful for diagnostic purpose.

Clear cell meningioma can be confused with other clear cell tumors or clear cell-like tumors, for example renal cell carcinoma, chordoma, hemangiopericytoma, chondroma or untypical osteosarcoma[1,3]. Lee et al. reported the magnetic resonance imaging features of clear cell meningioma[13]. However, it appears to be difficult to make a correct diagnosis by clinical fingings alone.

The biologic behavior of meningiomas is variable. Clinical prognosis is correlated with histopathologic parameters[3,14,15]. Zorludemir et al. reported that recurrence was noted in 61% of cases, local discontinuous spread in 15%, widespread cranial to spinal metastasis in 8% and mortality in 23%[7,16]. The present case has not reffered any recurrence in the 2 years since surgery. However a much longer follow-up will be necessary with this patient.

Younger patients undergoing multiple laminectomy are reported to incur kyphotic change at involving the spine within 5 years in 3 to 7 % of the cases[17]. In order to obviate the possibility of such deformity, we performed en block laminectomy with T-Saw and replaced the resected laminas in order to reconstruct the posterior element of the spine after resection of tumor. It is now 2 years since this surgery was undertaken and this case has not, so far, exhibited any deformity.

In summary, we reported a rare case of spinal clear cell meningioma in 10-year-old girl. It was characterized at the point of younger age and multiple involvement of lumbar spine clinically. Correct diagnosis was attained with pathological examination characterized by rich glycogen in cytosol and ultrastructure of meningioma. Additionally, we performed en block laminectomy with T-Saw and replaced the resected laminas to reconstruct posterior element of the spine to obviate the possible kyphotic deformity.

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