Clear-cell meningioma of the cauda equina

Abstract  Meningiomas are rare tumours in children and lumbar lesions are exceptional. We report a clear-cell meningioma (CCM) of the cauda equina in a 10-year-old girl. The tumour was diagnosed by MRI, showing an enhancing intradural mass extending from L1 to L4. Pathology and immunohistochemical study demonstrated a CCM. The patient had a recurrence 6 months after the operation requiring further surgery. CCM are rare lesions, characterised by abundant cytoplasmic glycogen particles. Complete surgical removal is necessary because, despite their benign histological appearance, CCM are potentially aggressive and may recur, spread locally and even metastasize.

Key words  Meningioma · Spine · Magnetic resonance imaging

Introduction

Intraspinal meningiomas are rare in children, particularly in the lumbar region. We report a case with unusual histology.

Case report

A 10-year-old girl had a 3-year history of backache, developing gradually, associated with cramps in the legs. She also had a painful scoliosis but examination was otherwise normal. When she was admitted for episodic back pain and bilateral leg weakness, examination revealed a bilateral motor and sensory deficit with a T10 level. There were no cutaneous stigmata.

MRI showed an intradural extramedullary mass filling the spinal canal from L1 to L4 (Fig. 1). The mass was isointense with the spinal cord on T1- and T2-weighted sequences and enhanced homogeneously except in its central part, which remained of low signal and was thought to represent necrosis. The conus medullaris was displaced posteriorly. Delineation of the mass and the cauda equina was difficult. A mild degree of spinal canal enlargement from T12 to L5 was noted. The preoperative diagnosis was intradural ependymoma or schwannoma.

At surgery an encapsulated tumour was found. No firm connection with dura mater or cauda equina was found at its lower part, but tight adhesions to nerve roots were found at its upper pole, near the conus medullaris. These were easily stripped after section of a few rootlets. Gross total resection of the tumour was achieved.

Peroperative frozen sections did not permit a definite diagnosis between ependymoma and meningioma. Gross pathology showed a round, well-circumscribed mass and, on light microscopy, the tumour showed conspicuous fibrosis and a rich vascular network. The tumour cells had a rather large, clear cytoplasm and round, regular nuclei without mitoses. They were organized in nests with a few small whorls. These appearances were more in favour of a meningioma than of a clear-cell ependymoma. Immunohistochemistry demonstrated staining with vimentin and neurone-specific enolase (NSE) and questionably with S-100 protein. Chromogranin, glial fibrillary acid protein (GFAP) and epithelial membrane antigen (EMA) were negative. These results confirmed the diagnosis of clear-cell meningioma (CCM) (Fig. 2).

Postoperative recovery was uneventful except for a mild deficit and slight wasting of the left leg. One month and a half later, MRI revealed no obvious residual tumour, except for a small enhancing

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Abstract  Meningiomas are rare tumours in children and lumbar lesions are exceptional. We report a clear-cell meningioma (CCM) of the cauda equina in a 10-year-old girl. The tumour was diagnosed by MRI, showing an enhancing intradural mass extending from L1 to L4. Pathology and immunohistochemical study demonstrated a CCM. The patient had a recurrence 6 months after the operation requiring further surgery. CCM are rare lesions, characterised by abundant cytoplasmic glycogen particles. Complete surgical removal is necessary because, despite their benign histological appearance, CCM are potentially aggressive and may recur, spread locally and even metastasize.

Key words  Meningioma · Spine · Magnetic resonance imaging
Fig. 1a–c  Sagittal MRI. a T2-weighted and b, c T1-weighted images before and after contrast medium show a well-defined mass displacing the conus medullaris posteriorly; the cauda equina is embedded in the mass, which is isointense with the spinal cord in a, b. Marked and homogeneous contrast enhancement is seen, except in its central part. Note mild spinal canal enlargement.

Fig. 2a, b  Pathology specimen. a Haematoxylin and eosin (HE) original magnification × 16; b original magnification × 40. Clear cells, with normal nuclei, are organised in whorls and interspersed with hyaline sheets.