CASE REPORT

Primary spinal cord desmoplastic astrocytoma in an adolescent: a rare tumour at rare site and rare age

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We report a case of a non-infantile primary intramedullary spinal cord desmoplastic astrocytoma in an 18-year-old girl who presented with spastic paraparesis. The patient had been unable to run for 1 year. Magnetic resonance imaging of her spine showed an intramedullary solid and cystic heterogeneously enhancing lesion located at T7-T8 level. Partial excision was performed. Histology revealed a desmoplastic astrocytoma. To the best of our knowledge, there is no report on primary desmoplastic astrocytoma of the spinal cord in literature. Nor has such a symptomatic tumour manifesting at the age of 18 years been documented.

Introduction

Desmoplastic infantile astrocytoma (DIA) is a rare supratentorial tumour, usually reported in the first 24 months of life; the mean age at presentation being around 5 to 6 months. The non-infantile DIA is a rare variant; in only six instances has it been reported in older children.[1-5] In two of these patients, clinical symptoms (seizures) ensued at the age of 3 and 6 months, though the histological diagnosis was made at the age of 7 and 9 years, respectively,[1,2] and the other four cases were reported in patients aged 3.5 to 11 years. In addition to the intracranial involvement, one of these patients also had metastases/multifocal lesions involving the spinal cord at the presentation.[3] However, the patient's spinal lesions remained untreated and histological proof of their nature was never obtained.

Case report

An 18-year-old girl presented with gradual bilateral lower limb weakness and stiffness for 2 years. She had been unable to run for 1 year. Her symptoms had increased over the last 2 months during which time she was experiencing frequent falls. On examination she had spastic paraparesis, with lower limb power at 2/5 on the right and 3/5 on the left, upgoing plantar reflexes and hyper-reflexia. Tendon reflexes in the upper limbs were brisk, but her cranial nerves were intact. Urgent magnetic resonance imaging (MRI) of the spinal cord showed an intramedullary solid and cystic heterogeneously enhancing lesion located at the T7 to T8 level. The solid component was predominantly T1-weighted (T1W) and T2-weighted (T2W) isointense with a smaller T1W iso-to-hypo-intense and T2W hyperintense cystic component at its caudal end. There was an associated syrinx extending from T6 to T9 levels (Figs 1 and 2). The MRI of her brain was entirely unremarkable. In view of the difficult resection, partial excision with debulking was performed. Histopathology showed an infiltrative tumour in a collagenous stroma. Tumour cells were round to fibrillary; some cells occurred in the form of broader sheets and clusters, but there were no neuronal cells. No mitosis, necrosis, or endothelial proliferation was seen. Tumour cells were positive for glial fibrillary acidic protein and equivocal for cytokeratin and epithelial membrane antigen. There was an MIB1 labelling index of around 3%. Overall features were suggestive of desmoplastic astrocytoma. The postoperative period was uneventful and the patient can now walk with stick, and there is no sphincter disturbance. Follow-up imaging over a period of 4 years showed small static residual component.

Discussion

Desmoplastic infantile astrocytomas usually present as superficial cerebral masses attached to the dura. Usually they are large slow-growing tumours, and often have a good prognosis after gross surgical resection alone. Radiologically, intracranial DIAs manifest as broad dural-based cystic masses with peripheral solid-enhancing components.

Key words
Adolescent; Astrocytoma; Spinal cord

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一名青年的原發性脊髓促結締組織增生型星形細胞瘤：罕見的腫瘤、罕見的位置和罕有的病發年齡

本文報告一名18歲少女患有非嬰兒原發性髓內促結締組織增生型星形細胞瘤，病發時出現痙攣性輕截癱。患者已有一年未能跑步。脊柱磁共振影像顯示在T7-T8髓內有囊實性不均勻增強型病灶。患者接受部分切除術，組織學顯示促結締組織增生型星形細胞瘤。據我們所知，文獻中並未有記載原發性促結締組織增生型星形細胞瘤發生在脊髓的位置，亦未有文獻報告這種具症狀的腫瘤發生在剛成年的患者身上。

In the brain, the differential diagnosis includes desmoplastic infantile ganglioglioma, pleomorphic xanthoastrocytoma, low-grade astrocytoma, glioblastoma, oligodendroglioma, and primitive neuroectodermal tumour.

Common intramedullary spinal cord neoplasms in children and young adults include astrocytoma,

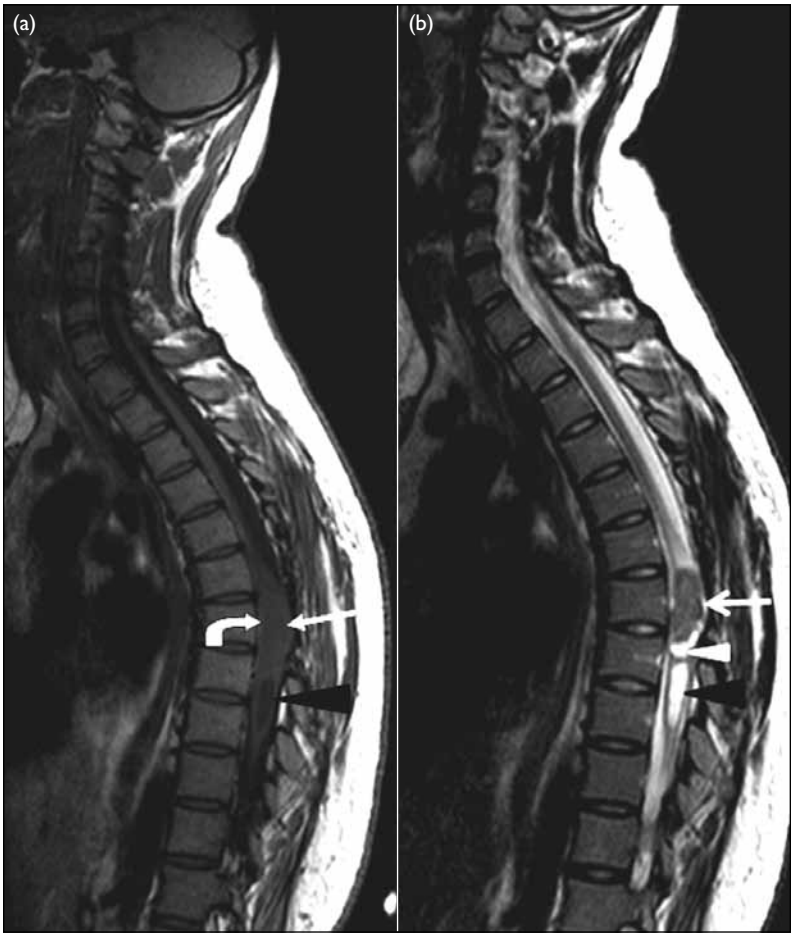

FIG 1. (a) T1-weighted sagittal image of spine showing intramedullary hypo-to-iso-intense lesion extending over T6 and T7 vertebral bodies (white arrow). Note the hyperintense rim (curved arrow) and syrinx (black arrowhead). (b) T2-weighted sagittal image of spine showing iso-to-hyper-intense intramedullary extending over T6 and T7 vertebral bodies (white arrow). Note the small hyperintense cystic components (white arrowhead) and syrinx (black arrowhead)

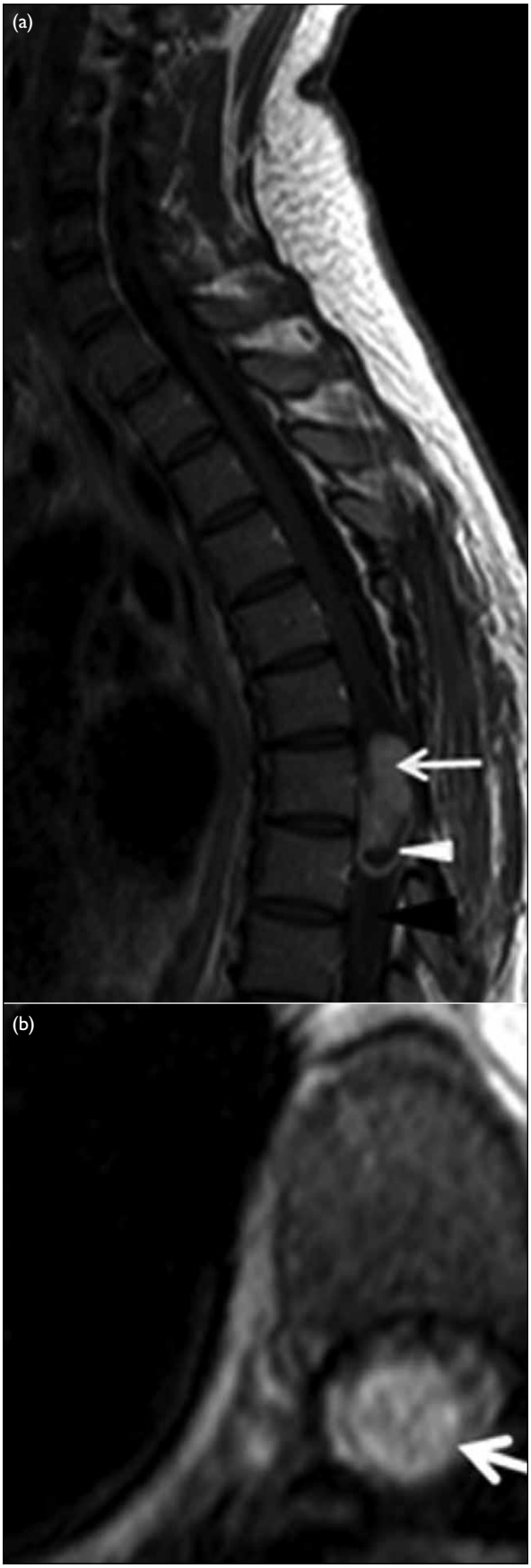

FIG 2. T1-weighted post-contrast (a) sagittal and (b) axial images of spine showing heterogeneous enhancement of the lesion (white arrows). Note the small inferior rim-enhancing cystic component (white arrowhead) and syrinx (black arrowhead)

ganglioglioma, and ependymoma. The imaging modality of choice is MRI, which can identify an intramedullary spinal cord neoplasm. Due to overlapping features with presence of solid-cystic components, heterogeneous enhancement, associated cysts and syrinx, desmoplastic astrocytomas of the spinal cord remain indistinguishable from the other intramedullary spinal cord lesions at this age-group. At young age, astrocytomas and gangliogliomas are generally commoner than ependymomas and often reveal heterogeneous eccentric enhancement causing an asymmetric enlargement of the spinal cord. On the other hand, ependymomas tend to reveal homogenous centrally located enhancement, and are often associated with rostral and caudal cysts. Although MRI does not provide a histological diagnosis of the lesion, its location, exact extent, and mass effect can reliably predict its nature.

In conclusion, non-infantile desmoplastic astrocytoma is a rare entity. To our knowledge, primary desmoplastic astrocytoma of the spinal cord has never been reported in literature. Moreover, the age of presentation (18 years) of a non-infantile desmoplastic astrocytoma also appears unique. Although the final diagnosis always relies on histology, imaging is crucial in evaluation of its exact extent, size and location, which can facilitate better surgical management and helpful in planning follow-up imaging to look for residual/recurrence of the disease.

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