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LETTER TO THE EDITOR

Rosette-forming glioneuronal tumour of dorsolumbar spinal cord

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A 14-year-old boy presented to our clinic with history of gradually progressive paraparesis for the last 10 years. The patient also had history of urinary incontinence and constipation. Both lower limbs were spastic (Ashworth grade3) on examination and motor power was 0/5 (MRC grade) in both lower limbs.

His magnetic resonance image showed a contrast enhancing lesion extending from D7-L1 (Figs.1 and 2). The lesion was ill-defined and compressing the spinal cord to the right (axial images, Fig. 3). The patient also had scoliosis of the dorsolumbar spine (X-ray image, Fig. 4) with primary curve to the right and compensatory bend to the left. He underwent D6-L2 exploration and decompression of the tumour. Intraoperatively, a greyish white soft suckable moderately vascular tumour was identified with poor plane from the surrounding cord. The histopathological examination of the tumour revealed a rosette-forming glioneuronal tumour (RGNT). Microscopically, the tumour was composed of monomorphic and bland cells arranged in repetitive small rosettes (Fig. 4). A 6-month follow-up of this patient showed marginal improvement in power to MRC grade $^{2}/_{5}$ and reduction in spasticity (Ashworth grade 2).

Cases of similar rosette-forming glioneuronal tumour have been reported involving the posterior fossa, pineal region, optic nerve, hypothalamus and cervical spinal cord. The posterior fossa tumour was composed of bland neurocytes arranged in rosettes along with glial areas resembling pilocytic astrocytoma. MIB-1 labelling index was reported < 1% [2, 5]. There was no necrosis, anaplasia nor microvascular proliferation. The neurocytic rich region showed strong synaptophysin positivity. The surrounding glial tissue showed GFAP positivity.

Till date, only one patient has been reported to harbour a dorsolumbar (D7-L1) RGNT. This 27-year-old female presented with paraplegia and bladder-bowel incontinence. Radiological evaluation showed a T1 hypointense and T2 hyperintense lesion with heterogenous contrast enhancement. Following a gross total excision of the lesion, she has remained clinically stable at 15-month follow-up duration [4].

Discussion

RGNT is a new pathological entity with only about 50 cases reported in literature. Female preponderance has been recorded and mean age is of around 31 years. A single case report of patient with NF1 harbouring RGNT in the optic nerve has been described. Radiological appearance of RGNT comprises of T1 iso-to hypointense lesion and T2 hyperintense lesion with occasional presence of calcification. This lesion usually appears well circumscribed and comprises solid and cystic areas on MR images. Rare satellite lesions have also been described.

Only five cases of spinal RGNT have been reported till date. Pathologically, these lesions are soft, gelatinous well-circumscribed areas with minimal surrounding region of infiltration. On microscopy, two separate components have been described [1]. The neurocytic portion has uniformly arranged rosettes of neurocytes and perivascular pseudorosettes. The glial component usually resembles pilocytic astrocytoma with spindle and piloid

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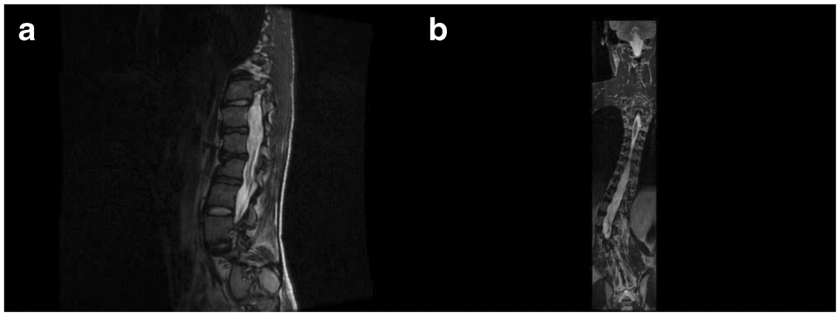

Fig. 1 **a** T2-weighted sagittal image. **b** T2-weighted coronal image

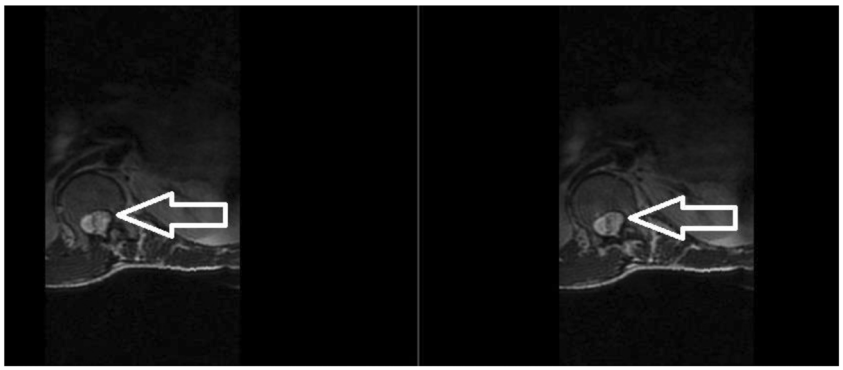

Fig. 2 Contrast-enhanced axial MR image (white arrow pointing towards the lesion compressing the cord)

cells. Thrombosed vessels and rosenthal fibres are rarely seen. These are classically low-grade tumours with infrequent cytological atypia and mitotic activity [3].

The fourth ventricular glioneuronal tumour typically affects young adults and is a low-grade lesion [6]. It was first included in the WHO classification in 2007. In the 2016 modification of WHO classification, these were renamed as rosette-forming glioneuronal tumours. A common morphological feature of RGNTs involving the fourth ventricle, spinal cord, third or lateral ventricles or septum pellucidum is the presence of ependymal covering.

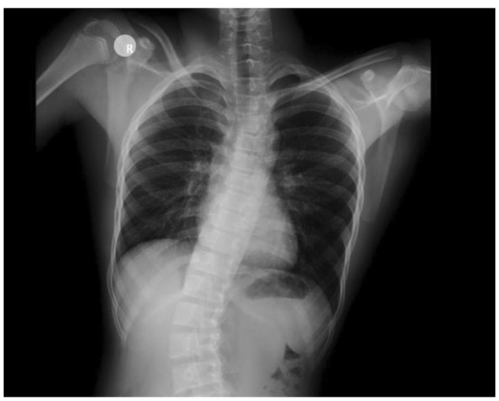

Fig. 3 X-ray (chest and abdomen) AP view

Surgery is the preferred modality of treatment. Radiotherapy or adjuvant chemotherapy is not recommended for these low-grade lesions as they usually take a benign clinical course.

Spinal RGNTs pose a significant diagnostic dilemma as they resemble intramedullary tumours like ependymomas and astrocytomas. Ependymomas tend to be located in the upper cervical or thoracic spinal cord and more commonly affect young adults (age < 30 years). These usually have a good demarcation from the surrounding normal spinal cord and are located centrally. These present with heterogenous enhancement on the post gadolinium MR imaging. Astrocytomas commonly affect teenagers and present as homogenous contrast-enhancing lesions. They usually infiltrate into the surrounding parenchyma and are less well demarcated. Syrinx formation was more frequently associated with ependymomas than astrocytomas. While in ependymoma, the pathological picture demonstrated papillary arrangement, perivascular pseudorosettes and ependymal rosettes. Perivascular tumour showed GFAP, S100 and vimentin positivity and being negative for Syn and NSE. Astrocytomas on the other hand comprised of densely woven tumour tissue alternating with loose connective tissue and occasional cystic change and Rosenthal fibres with S100, GFAP and vimentin positivity.

We want to apprise the neurosurgical community with this new and rare pathological entity. These tumours can lead to severe neurological deterioration, decreased

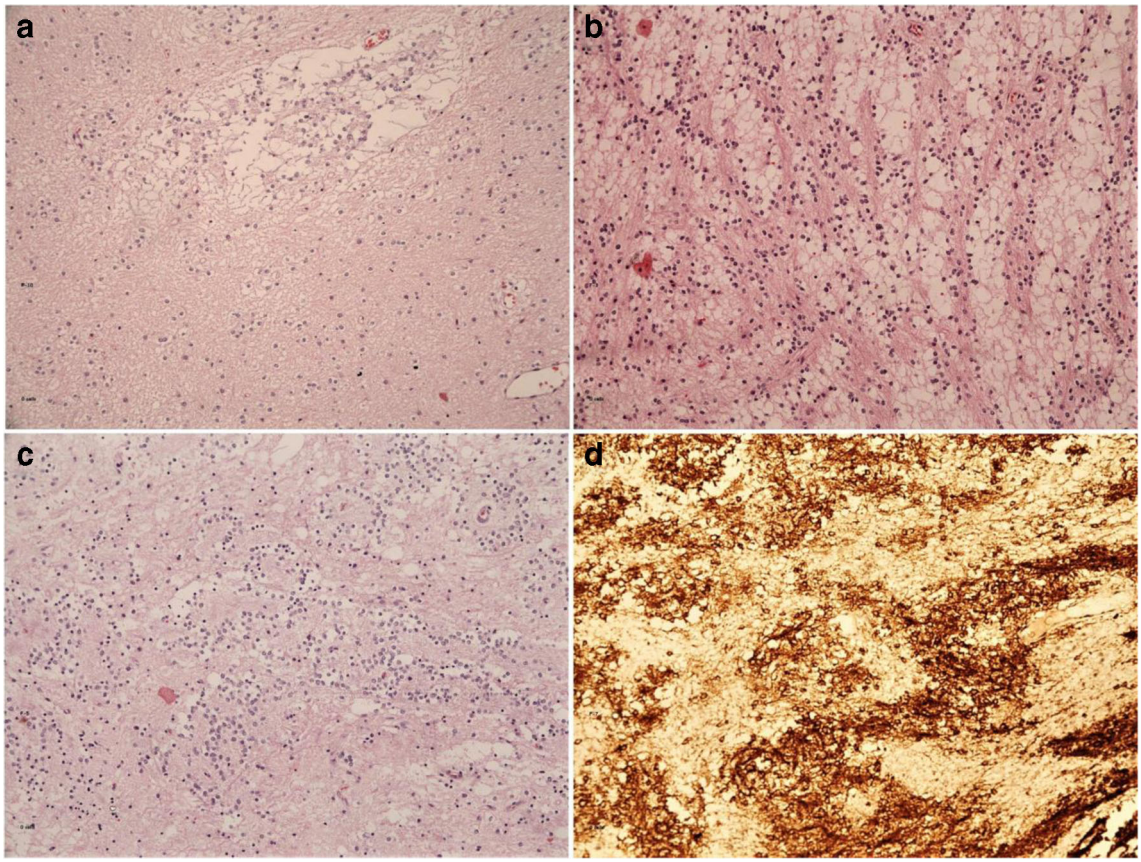

Fig. 4 Histopathological imaging. **a**, **b** Light microscopy showing fibrillary tumour with low cellularity and microcystic change. **c** The tumour cells are monomorphic and bland, arranged in repetitive and small rosettes. **d** The immunostaining for neuronal marker MAP2 shows neuronal differentiation in the tumour cells forming rosettes

functional activity and poor quality of life if not detected at an early stage. Gross total resection followed by close clinical follow-up and genetic analysis could help estimate prognosis better in these cases. These tumours behave in a benign fashion, though the current clinical experience with spinal RGNT is not very long.

Compliance with ethical standards

Conflict of interest None.

Statement of informed consent Written/informed consent was obtained from the parents of the patient for utilising the clinical details and images for journal submission and it has been stated that no personal/contact information of the patient/ parents will be shared with anyone.

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