A Case of Tethered Cord Associated with Amniotic Band Syndrome

Sir,

Amniotic band syndrome is a set of congenital malformations ranging from minor constriction rings of the digits to complex, multiple congenital anomalies that are attributed to amniotic bands that stick, entangle and disrupt fetal parts. 1 Neural tube defects, heterotropias, acalvaria and many other cranio-facial anomalies are reported with this syndrome. 2 We report a case of spinal dysraphism with cervical cord tethering associated with Amniotic band syndrome.

A 27 year old, pure vegetarian presented with history of numbness of distal lower limbs of 3 months duration. Examination showed a constriction band on right arm with limb below the band atrophic (Fig. 1A- arrow), a subcutaneous swelling over the posterior neck and absent skull bones over fronto-parietal region (only skin and subcutaneous tissue covering the scalp). All these were present from birth. Neurological examination and investigations suggested a diagnosis of possible vitamin B12 deficiency. MRI cervical cord and brain didn’t show any features of B12 deficiency, but it showed some incidental findings:- 1) A fibrous strand connecting the upper cervical cord (Fig. 1B - T1 weighted image- white arrow) to a mixed signal intensity swelling (?lipoma) on the posterior part of neck (Fig. 1B- black arrow), with tethering of cord and a small syrinx. No posterior arch of atlas seen (spinal dysraphism). 2) Cisterna magna is large and freely communicates with fourth ventricle. 3) Tentorium cerebelli is absent (filled with CSF) (Fig. 1C-T2 weighted image- arrow). 4) Nodular heterotropia near the right lateral ventricle. 5) Large calvarial defect in parietal bone. As the MRI findings are not associated with any symptoms, patient was started on B12 injections and discharged.

Our patient presented with features of neuropathy, possibly due to B12 deficiency. The constellation of incidental clinical and MRI findings in our patient with a congenital constriction band, is suggestive of a diagnosis of Amniotic band syndrome. 3 All the anomalies seen in this patient, except absent tentorium cerebelli are described with the syndrome. To our best knowledge, spinal dysraphism with cervical cord tethering and a number of associated anomalies in a single patient of amniotic band syndrome, are not reported so far.

In conclusion, we report a rare and interesting case of amniotic band syndrome with cervical cord tethering (spinal dysraphism) and a number of other associated anomalies.

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Fig. 1A : Shows a constriction band on right arm (black arrow) of the patient.
Fig. 1B : (T1 weighted image) shows a fibrous strand connecting cervical cord (white arrow) to a mixed signal intensity swelling on the posterior part of neck (black arrow), with tethering of cord and a small syrinx.
Fig. 1C : (T2 weighted image) shows tentorium cerebelli is absent and filled with CSF (black arrow).