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Letter to the Editor

A rare case of giant multiseptated thoracic myelomeningocele with segmental placode

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Sir,

Myelomeningocele is a common form of spinal dysraphism involving thoraco-lumbar region. Pure thoracic form is quite rare. We present a unique case of giant, multiseptated thoracic myelomeningocele with a segmental placode, both proximal and distal to which the spinal cord was normally neurulated and discuss the unique features associated with such rare variety of thoracic myelomeningocele.

A 16-month-old male child presented with huge swelling over thoracic region since birth, which was progressively increasing in size. The parents did seek for the neurosurgical consultation and were advised surgery soon after the birth but they deferred it for lack of money and risk for further neurological deficit. There was no history of leakage of fluid from the swelling. The swelling was approximately 15 cm × 12 cm × 12 cm in size, lobulated, with broad pedicle of skin, and covered with thick desquamated epithelium [Figure 1a-c]. The child was moving his both legs normally and was able to stand with support. There was no obvious kyphosis or limb deformity. Sensation was apparently normal with the child responding to painful and touch stimuli to both lower limbs. The child was voiding urine spontaneously and intermittently with no continuous dribbling or history suggestive of recurrent urinary tract infections. Childs' head size was large with circumference being 56 cm (approximately 22 inches) with anterior fontanelle still open. Magnetic resonance imaging of thoracic region showed a large, multiseptated, swelling whose most of the part was T2 hyperintense suggestive of cerebrospinal fluid (CSF) collection and a part of thoracic cord entering into the neck of swelling [Figure 2]. There was a long stalk extending from the neck of swelling to fundus region. Computed tomography brain showed presence of hydrocephalus. A right sided low pressure ventriculoperitoneal shunt was done to prevent the chances of postoperative CSF leak. The dissection done all around the myelomeningocele sac at its junction with the normal skin. The dural covering was intact around maximum part of the swelling like a nearly closed neural tube except for a narrow midline gap. The swelling was connected to normal dura through a large pedicle [Figure 3]. The swelling was opened at its fundus region. There was a fibro vascular stalk connecting the fundus of swelling to the spinal cord, causing tethering of the latter.

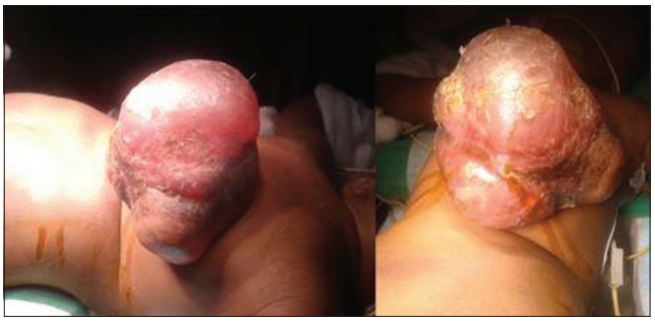

Figure 1: (a) Large, lobulated swelling in thoracic region covered by intact skin in its base and thick desquamated epithelium at top (b) Thoracic swelling from caudal view

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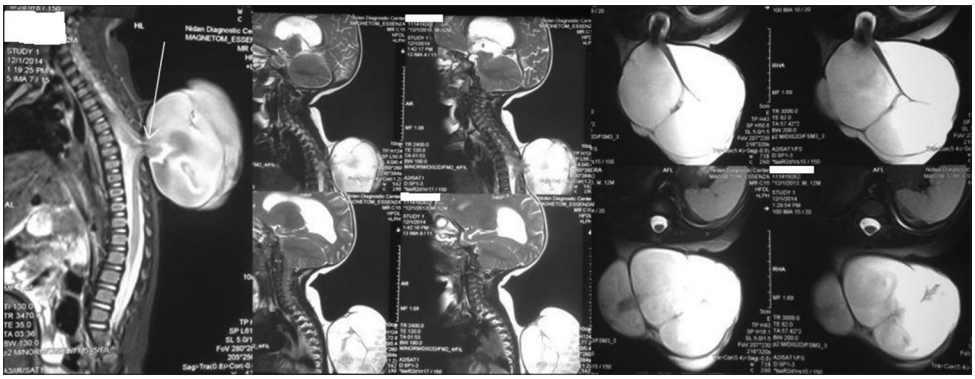

Figure 2: Magnetic resonance imaging thoracic region shows a small neural placode at the base of swelling (arrow) with a stalk connecting the placode with dural covering. Multiseptated nature of the swelling can be seen along with hydrocephalus

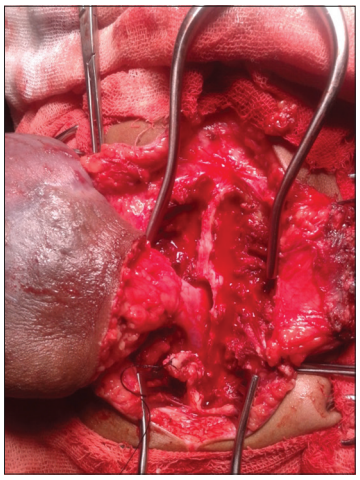

Figure 3: The large swelling connected to the normal dura by a broad pedicle

The stalk was excised along with the redundant dural sac. A small neural placode seen at the base of swelling with the normal cord distal to the placode. The pial layer of two sides was sutured in the midline. Dura was closed primarily. Multilayered closure technique was adopted for closing the muscle, thoraco-lumbar fascia, subcutaneous tissue, and skin. Postoperative course was nonsignificant with only superficial infection of upper part of suture line which was managed conservatively. No deterioration of neurological function occurred.

Giant myelomeningoceles are challenge due to high-risk of skin breakdown, CSF leak, infection, etc., To reduce the chances of skin breakdown and subsequent CSF leak, a ventriculoperitoneal is advisable such cases. We prefer a two sitting shunt and defect repair to decrease the chances of shunt infection as we did in this case. We performed a low pressure shunt as the anterior fontanelle was open and there was no radiological signs of grossly increased intracranial pressure (periventricular lucency). In presence of gross neurologic deficits in thoracic myelomeningocele, there occurs invariably kyphosis due to unrestricted pull of the normally innervated proximal anterior abdominal and intercostal muscles. This kyphosis prevents the tensionless closure of the defect and requires same sitting kyphectomy. Fortunately in our case, due to normal neurulation of distal cord the paraspinal muscles had retained their tone and this explains the lack of kyphosis in the present case. Segmental neural placode is extremely rare and account for only 4% of all open neural tube defects. Exact mechanism of genesis of such segmental placode is not clearly known but a "square-pulse" type teratogenic insult during primary neurulation has been proposed which results in normal neurulation starting distal to an isolated failure of neural plate fusion. "Collision site" hypothesis explains the cause as failure from two adjacent neurulation sites proceeding in opposite directions.[3] The most common site of a segmental placode is midthoracic or thoraco-lumbar. The secondary neurulation process remains unaffected with normal closure of posterior neuropore and formation of conus. The presence of normal or near normal neurological functions in the lower limbs in presence of a high neural tube defect such as thoracic or cervical variety should raise the suspicion of a segmental placode.

Thoracic and cervical region myelomeningoceles are quite rare and occur in 1–5% of all neural tube closure defects.[4] They are usually associated with a placode that sits at the base of the swelling rather than floating on the top of it. Tethering by a fibro neurovascular stalk is common as in the present case and needs detethering to prevent future neurological deterioration. They are usually associated with less neurological deficits than their lumbosacral counterparts[2,5] but if left untreated, they can lead to progressive neurologic deficit due to tethering. Careful intradural exploration and microsurgical release of the spinal cord by meticulous resection of all tethering bands has been suggested.[1] The present case tries to heighten the awareness of such unique presentation of the myelomeningocele in the thoracic spine.

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Conflicts of interest
There are no conflicts of interest.

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