

A rare case of giant multiseptated thoracic myelomeningocele with segmental placode: Commentary

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Sir,

It is with great interest that we read the paper entitled, “A rare case of giant multiseptated thoracic myelomeningocele with segmental placode” by Patnaik and Mahapatra.^[4] The authors described a rare presentation of a thoracic spinal dysraphism. The child they presented had no neurological and urologic deterioration, which is very uncommon in a thoracic myelomeningocele, and the magnetic resonance imaging (MRI) showed a multiloculated swelling with a fibroneural stalk that linked the skin malformation to the underlying spinal cord.

However, the complete description of the malformation and the fine MRI images and peroperative pictures provided by the authors may not be consistent with the diagnosis of a true myelomeningocele. We believed that it is a form of spinal dysraphism called “limited dorsal myeloschisis” (LDM). Such entity first described in 1993^[1] has been nicely reviewed by Pang *et al.* with its complete description^[2] and classification.^[3] The case described by Patnaik and Mahapatra would correspond to a typical case of the saccular form. It has the skin-based sac which is filled with cerebrospinal fluid (CSF). The top of the skin lesion is recovered by desquamated tissue which is not normal skin. The neurologic condition is excellent in this case report as reported by Pang *et al.* in 40–50% of the thoracic and lumbar cases, where the urologic deterioration is only present in 15–20% of the cases.^[2] In the present report, MRI findings are also consistent with Pang’s description. The fibroneural stalk traverses the CSF-filled sac to reach the top of the skin anomaly. We can see on the axial view that there is a little depression at the junction between the top of the sac and the stalk that corresponds to the skin crater and presumably to the original site of the disjunction failure.^[2]

There is one very important point brought by the authors: The presence of an evolutive hydrocephalus that required a shunt.^[4] This is very interesting as Pang *et al.* did not report the presence of hydrocephalus in patients with thoracic LDM.^[2]

This distinction between LDM and myelomeningocele has also an embryological implication: The difference lying in the degree of the neurulation stage.^[2] In LDM, only the final stage of the primary neurulation is incomplete. Therefore, a limited disjunction between cutaneous and neural ectoderms is present. It prevents the complete midline skin closure (the top of the sac is covered by desquamated tissue which is not normal skin) and allows persistence of a physical link (fibroneural stalk) between the disjunction site and the dorsal neural tube. This is well documented by the peroperative pictures provided by the authors. The swelling was connected by this stalk to the dura trough a very narrow

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midline bone defect shown in Figure 3 from Patnaik and Mahapatra.^[4]

The report from Patnaik and Mahapatra^[4] underlines the complexity in the description and classification of spinal dysraphisms. Because these LDMs and myelomeningoceles have such great difference both in the natural history and final outcome, especially at the thoracic level, the proper distinction between these two entities is of paramount importance for a correct management of the child and a proper counseling to the parents.^[5]

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Conflicts of interest

There are no conflicts of interest.

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