Thoracic myelocystocephalocele in a neurologically intact infant

E. O. Komolafe, M. T. Shokunbi, and A. O. Malomo

Department of Neurosurgery, Unit of the Hospital for Surgery, University College Hospital, P. M. B. 5116, Ibadan, Nigeria.

E-mail: adeyosin2001@yahoo.com, adeyosin@comfe.edu.ng

Summary

Objectives: To report an uncommon and rare case of a high congenital spinal anomaly with an unusual presentation.

Methods: This is a case presentation with relevant radiological and operative findings.

Conclusion: This lesion is a differential diagnosis of cystic congenital spinal mass lesions. This case is an example of a high congenital spinal lesion with very minimal or negligible neurological deficits, with no other congenital malformations.

Keywords: Thoracic spine, Myelocystocephalocele, Intact nervous system.

Résultat

Rapporteur un cas peu commun et un cas rare d'une anomalie congenitale vertébrale très sévère avec une présentation peu commune, c'est un cas avec des résultats opératoires et radiologiques pertinents.

Cette lésion est un diagnostic différentiel de masse lésions Kyste congenital vertébral. Ce cas est un exemple d'une lésion spinale congenitale bien élevée avec des déficits neurologiques très minimes ou négligeables avec aucune malformation congenitale.

Introduction

Congenital malformations of the nervous system occur commonly worldwide, and spinal dysraphism is the most common. These spinal malformations can occur as isolate lesions or as part of a large complex midline developmental abnormality.

Myelocystocele and myelocystocephalocele are however very rare spinal congenital lesions. Usually the higher the spinal anomaly is situated, the worse the neurological status except in the straightforward meningococele. A case of myelocystocephalocele of the thoracic spine in a neurologically intact infant is presented in this communication.

Case report

AA, a male neonate and second in the family presented to our neurosurgical service on 23/9/98, on the eighth day of life with the history of a midline, mid-thoracic cystic mass lesion noticed at birth (fig. 1). There has been no change in the size of the mass since birth. There is associated grunting respiration aggravated at times by sleep. There was no family history of similar or any other anomalies. There was also no history suggestive of urinary or faecal incontinence. The pregnancy and delivery were uneventful.

Examination revealed a pink and active male neonate

Fig. 1 A neonate with an oval mid-thoracic cystic mass

Fig. 2 CT myelogram showing contrast filled spinal mass

with stridor. The occipito-frontal circumference was 34.5cm (normal size is 34 +/-2cm). The anterior fontanelle was flat and aormotensive. There was good visual fixation and tracking to light. The spinal mass measured 7.5cm in its largest diameter and was covered with thin skin. There was mild

Correspondence

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spasticity of both legs with no limb or ankle deformity.

A clinical assessment of a mid-thoracic spina bifida cystica with a mild spastic paraparesis was made to rule out early hydrocephalus and Arnold-Chiari-malformation. Transfontanelle ultrasonography study was normal. The abdominopelvic ultrasound showed bilateral normal kidneys and urinary tract. The otolaryngological evaluation of the cause of the stridor was laryngotracheomalacia. This was clinically observed for sometime until the stridor improved indicating good cartilage deposition in the upper airway. CT Myelogram showed free flow of contrast between the spinal mass and the spinal subarachnoid space with prolapse of the cord into the soft tissue at T7 (fig. 2). The patient had surgery on 15/4/98. The findings were essentially as shown in fig. 3. A mid-thoracic oval cystic mass filled with cerebrospinal fluid (CSF) was found at surgery. Also a fistulous connection between the central canal of the cord and the cystic mass was seen (fig. 3). This was divided and excision of the mass performed with multiplier closure of the spinal defect.

He was discharged home a week later after an uneventful post-operative period. Neurological status however remained the same, but at the follow-up clinic of 8/6/98, the stridor was less in frequency and severity and the surgical wound scar was nicely healed. The occipito-frontal circumference was 43cm (expected size is 40+/-2cm) with flat and normotensive anterior fontanelle and the legs were now non-spastic. The findings of mild hypotonia, and spasticity of the limbs are not unusual in neurologically intact neonates as these resolve with time. A repeat transfontanelle ultrasound was reported to be normal. Further evaluation at subsequent follow up clinic attendance showed a neurologically intact child with no stridor and with excellent school performance.

**Discussion**

Myelocystomeningocele by definition is a skin-covered congenital cystic spinal lesion that is produced by an abnormal dilution of the central canal of the spinal cord with an associated meningocele. It is sometimes associated with extrophy of the cloaca, omphalocele and in 15% of patients with craniosynostosis. The aetiology and pathogenesis are unknown. It occurs sporadically in families. The recurrence risk for subsequent offspring of parents is negligible, usually less than 1%. Odekun in 1967 reported a case of lumbosacral meningocele without gross neurological deficit among the 96 cases of congenital spinal mass lesions.

This lesion is a differential diagnosis of cystic congenital spinal mass lesions. Other lesions such as oeunertic sinus with associated cutaneous haemangioma and a dermoid cyst within a meningocele, similar in location and presentation but with different pathology has been noted by other authors.

This case is an example of a high congenital spinal lesion with very minimal or negligible neurological deficits, without any other congenital malformations.

**References**