Thoracic myelocystomeningocele in a neurologically intact infant

***E. O. Komolafe[1], M. T. Shokunbi and A. O. Malomo**

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

[1]Department of Surgery, O. A. U. Teaching Hospital Complex, P. M. B. 5538, Ile-Ife, Osun State, Nigeria.

E-mail: adeyoyin2001@yahoo.com, adeyoyin@oauife.edu.ng

Summary

Objectives: **To report an uncommon and a 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, Myelocystomeningocele, Intact nervous system.***

Résumé

Rapporter un cas peu commun et un cas rare d'une anomalie congenitale vertébrale très sevère avec une présentation peu commune; c'est un cas avec des résultats operatoires et radioloques pertinents.

Cette lésion est un diagnostic différentiel de masse lésions Kyste congénital vertebral. Ce cas est un exemple d'une lésion spinale congénitale bien élevée avec des déficits neurologiques très minimum ou négligéiable avec aucune malformation congénitale.

Introduction

Congenital malformations of the nervous system occur commonly worldwide, and spinal dysraphism is the most common.[1,2,3] These spinal malformations can occur as isolate lesions or as part of a large complex midline developmental abnormality.

Myelocystocele and myelocystomeningocele are however very rare spinal congenital lesions. Usually the higher the spinal anomaly is situated, the worse the neurological status except in the straightforward meningocele. A case of myelocystomeningocele of the thoraic 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 2/3/98, 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 with stridor. The occipito-frontal circumstance was 34.5cm (normal size is 34 +/-2cm). The anterior fontanelle was flat and normotensive. 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

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

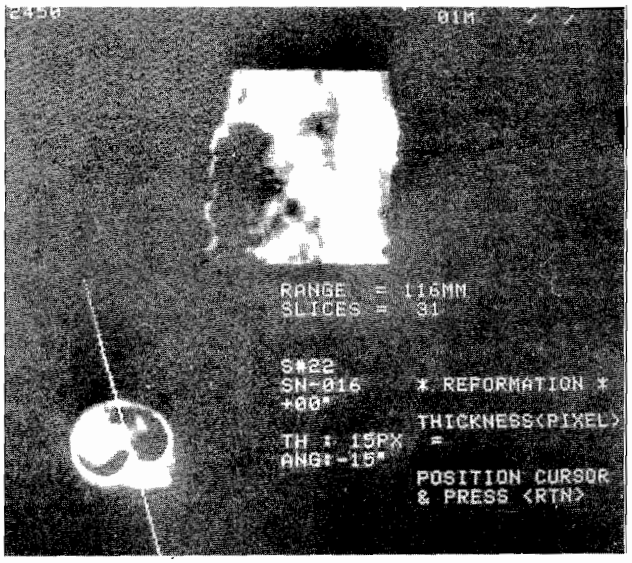

Fig. 2 *CT myelogram showing contrast filled spinal mass*

* *Correspondence*

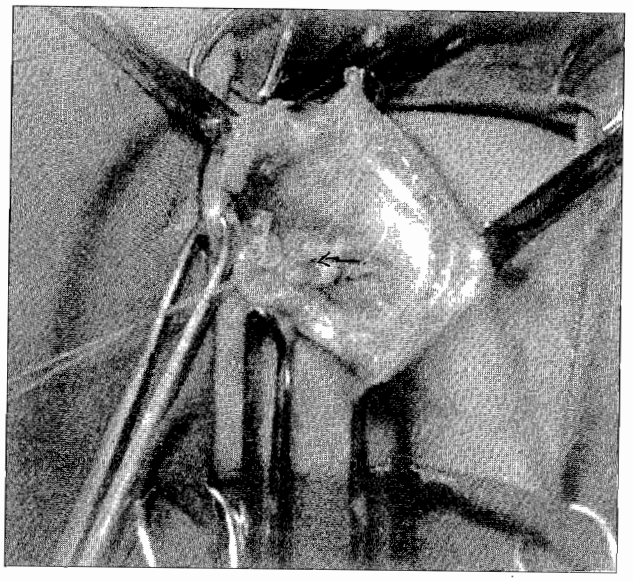

Fig. 3 ***Operating finding showing the connection between the central canal and the cystic mass.***

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.[2, 4, 5] Transfontanelle ultrasonography study was normal. The abdomino-pelvic ultrasound showed bilateral normal kidneys and urinary tract. The otorhinolaryngological 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 multiplayer 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 circumferences was 43cm (expected size is 40+/-2cm) with flat and normotensive anterior frontanelle and the legs were now non-spastic. The findings of mild hypertonia, 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 dilation 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 craniosynstosis.[6] 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%.[6]

Odeku in 1967 reported a case of lumbosacral meningomyelocystocele with gross neurological deficit among the 96 cases of congenital spinal mass lesions.[7]

This lesion is a differential diagnosis of cystic congenital spinal mass lesions. Other lesions such as neurenteric sinus with an associated cutaneous haemangioma and a dermoid cyst within a meningocele, similar in location and presentations but with different pathology has been noted by other authors.[8, 9]

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

References

1. Sarwark, JF: Spina Bifida (review). Paediatric clinic of North America 1996; 43: 1151 - 8.
2. Adeloye A: Congenital malformation of the cranio-spinal axis in Ibadan, Nigeria. Z. Kinderchir 1980; 31: 327 - 331.
3. Gupta B: Incidence of Congenital malformations in Nigerian children. West African Medical journal. 1996; 18: 22.
4. Rosenfeld JV and Watters DAK: Hydrocephalus and congenital abnormalities. In: Neurosurgery in the tropics - a practical approach to common problems. Macmillian education LTD. London, pp 126 - 132.
5. Vandertop WP, Asai A and Hoffman HJ et al: Surgical decompression for symptomatic chiari II malformations in neonates with myelomeningocele. J Neurosurg. 1992; 77: 541 - 544.
6. David DW: Genetics of development defects. In: Wilkins RH, Rengachary SS (eds) Neurosurgery. MCGRAW-HILL New York. pp 3430 - 3431.
7. Odeku EL, Grant IH and Ikop AC: Congenital malformations of the cerebrospinal axis seen in Western Nigeria. The spinal meningoceles. International surgery. 1967; 47: 580 - 589.
8. Grieve JP, Wilkins PR and Marsh HT: Neuronteric sinus dorsal to the thoracic spine with an associated cutaneous haemongioma. Case report, British Journal of Neurosurgery. 1998; 12: 259-263.
9. Sakai, Sakamoto K, Kobayashi N and Iguchi H : Dermoid cyst within an upper thoraic meningocele surgical neurology. 1996; 49: 287 - 292.