

Cervical Myelomeningocele and Hydrocephalus Without Neurological Deficit: A Case Report

¹T. Olugbenga Odebode, ¹S. Ukpong Udoffa and ²A. Donald Nzeh

¹Division of Neurological Surgery, Department of Surgery, ²Department of Radiology,
University of Ilorin Teaching Hospital, Ilorin, Nigeria

Abstract: Cervical myelomeningocele is a rare dysraphic lesion which rarely presents with neurological deficit in affected infants. We report a 9-day-old female neonate with a posterior midline neck swelling and a large head noted at birth; that had a mid cervical myelomeningocele and hydrocephalus without neurological deficit. All the brain ventricles were dilated on ultrasound and the fluid filled neck swelling showed no evidence of Chiari malformation or cord tethering. These associations were also absent during spinal exploration, repair of myelomeningocele and ventriculo-peritoneal shunting. Except for a shunt revision necessitated by shunt blockade occurring 12 days after surgery, there has been no neurological deterioration since eighteen months of initial surgery. We conclude that an infant with cervical myelomeningocele and hydrocephalus does not always have a Chiari malformation and could present without neurological deficit. A prolonged follow-up is however paramount in the event that the child may subsequently develop neurological deterioration.

Key words: Cervical • myelomeningocele • hydrocephalus

INTRODUCTION

Myelomeningocele is the commonest congenital malformation of the central nervous system (CNS) worldwide [1] and in Nigeria, it is the commonest anomaly in the north [2] and middle belt [3] but second to hydrocephalus in the south [4] the lumbo-sacral lesion being the most common [2-4]. From previous reports; the cervical type is very rare constituting only 3.0% to 8.0% of spinal bifida cystica [5, 6]. The degree of neurological deficit in infants with myelomeningocele increases the further away the lesion is from the conus. However, cases of isolated cervical myelomeningocele are usually associated with limited or no neurological deficit, though subsequent neurological deterioration could be fairly common. In this communication, we report a neonate who presented with cervical myelomeningocele associated with hydrocephalus but without neurological deficit.

CASE REPORT

A 9-day-old female; product of an uneventful term pregnancy, labour and delivery, had a posterior neck swelling which was first noted at birth. She is the third



Fig. 1: A 9-day-old neonate with an oval midline low cervical cystic swelling measuring 5 cm x 4 cm x 3 cm and a large head. The sessile base is covered with full thickness skin

child of a family devoid of history of CNS malformation. There was a round, fluctuant, translucent cystic neck swelling measuring 5 cm x 4 cm x 3 cm in size. Its wide base was covered with full thickness skin while its dome comprised of a tough purplish membrane without an evidence of cerebrospinal fluid leakage (Fig. 1). The active and pink neonate had normal growth parameters



Fig. 2: Plain cervical spine radiograph showing a spina bifida defect and widening of interpedicular distances of C₄₋₇ vertebral bodies

including a head circumference of 35 cm. The anterior fontanel was however full and 4 cm wide in its coronal plane. She had spontaneous movements in all extremities without focal pyramidal or cranial nerve deficit. The anal sphincter tone was normal and there was spontaneous voiding. She fixed and tracked promptly to moving targets. A plain cervical spine X-ray revealed spinal bifida defect and widening of inter-pedicular distances of the C₄₋₇ vertebral bodies (Fig. 2), while a trans-fontanel ultrasound scan (TFUSS) and ultrasound of the neck swelling revealed dilated lateral, third and fourth ventricles without an evidence of a Chiari type II or III anomaly. The cost of a cranio-cervical magnetic resonance imaging or cervical metrizamide myelography was not affordable to the parents. Her packed cell volume was 38% and blood chemistry was normal.

A ventriculo-peritoneal shunting, using a medium pressure Chhabra "Slit n spring" hydrocephalus shunt system (G. Surgiwear; India) and a repair of the myelomeningocele were performed. The major findings at spinal exploration were a myelomeningocele with a narrow neck, a dural fistula traceable into the spinal canal via a C5 lamina defect through which whitish strands of nerve rootlets connect the dorsal cervical cord to a placode at the center of the dome. There was no split cord malformation, syringomyelia or myelomeningocystocele. The nerve rootlets were detached bluntly from their attachment to the placode and dropped into the spinal canal. The reflected dura was double breasted and a simple layered closure was achieved with ease. Twelve days after shunting she developed a catheter blockage necessitating a revision of the shunting procedure. She was discharged ten days after revision and has enjoyed a hitch free clinical status for more than eighteen months.

DISCUSSION

Myelomeningocele; the commonest anomaly of the cerebrospinal axis worldwide, is a complex malformation of the spinal cord, nerve roots, meninges, vertebral bodies and skin resulting from a failure of closure of the neural tube in the developing foetus during the first four weeks of gestation. Its distribution along the vertebral column [6] is as follows: thoracic 5%, thoracolumbar 26%, lumbar 26%, lumbosacral 10%; with cervical myelomeningocele (CMMC) being the least common (3%). In Nigeria, most reported series of CNS anomalies lacked a cervical myelomeningocele [2-4]. The current case is the first cervical type to be reported from our centre.

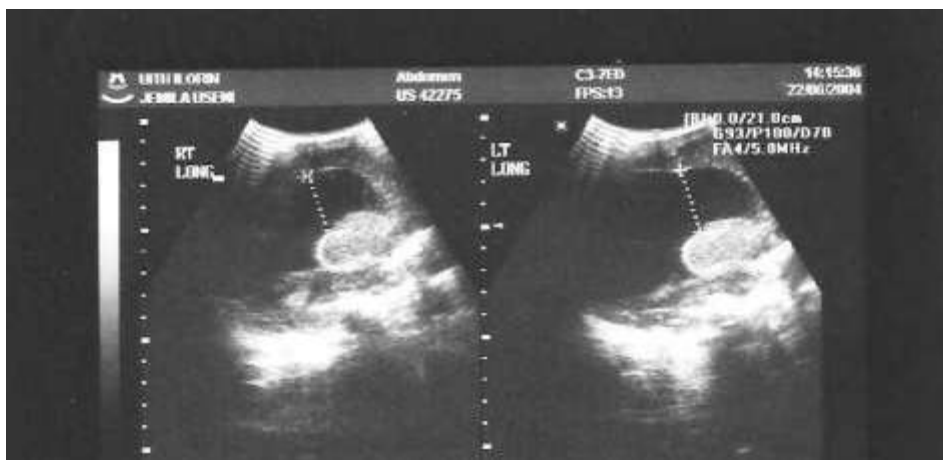


Fig. 3: Right and left parasagittal views of trans-fontanel ultrasound brain scan showing dilated right and left lateral ventricles due to hydrocephalus

The fact that neurological deficit worsens the higher a myelomeningocele lesion is above the conus appears to be restricted to patients with lumbo-sacral and thoracic dysraphic lesions. Patients with cervical lesions tend to retain normal neurology as in this and some other reported cases [5, 7]. In these patients the neural nodule most likely constitutes a vertical outgrowth from a functioning spinal cord in the form of a limited dorsal myeloschisis (LDM) and not a terminal placode as it obtains with thoracic and lumbosacral myelomeningocele [8]. Hence these patients manifest normal or near normal neurological function caudal to the level of the lesion whereas the patient with thoracic or lumbosacral lesion has no neurological function below the level of the terminal placode [8]. When a mild neurological deficit occurs, it may be due to an overlooked tethering of the cord resulting from a simple LDM in which the spinal cord is anchored to the base of the sac by a fibrovascular band [8]. In other infants, a myelocystocele may be responsible [7, 8]. On long term follow-up, a significant proportion of such infants experience progressive neurological deterioration due to untreated tethering, a condition which was notably absent in this case. For such patients, management should involve preoperative computed tomography or magnetic resonance imaging to identify tethering elements and at operation a single level laminectomy should be performed rostral to the dural stalk, exploration of the spinal canal and sac contents should be performed and fibrovascular stalk should be traced to their attachment with the dural surface of the cervical cord and then resected [8].

In the current patient, a diagnosis of cervical myelomeningocele (CMMC) is supported by a protuberant sac covered by a full thickness skin at its base and tough opaque squamous epithelium at the dome, distinguishing it from a myelocystomeningocele or a myelocystocele. In addition, the neural nodule was made up of a highly disorganized glioneural tissue and nerves on histology. A myelocystomeningocele is a skin-covered congenital cystic spinal lesion produced by an abnormal dilation of the central canal of the spinal cord with an associated meningocele [7, 9] representing a combined meningocele and focal hydromyelia and unlike CMMC, it tends to occur in association with omphalocele, bladder extrophy, imperforate anus and craniosynostosis [9]. A myelocystocele consists of an expanded ependyma lined cyst continuous with and therefore tethered to the underlying cervical hydromyelic cord. The combination of a cervical myelomeningocele and hydrocephalus in our

patient also raised the suspicion of a Chiari type II [10] or III [11] malformation, but this was debunked by findings at ultrasound and surgical exploration.

CONCLUSION

Cervical myelomeningocele associated with hydrocephalus could present without a neurological deficit.

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