CASE REPORT

Cervical Meningomyelocoele: A Case Report and Review of the Literature

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ABSTRACT

Cervical meningomyelocoele is a rare neural tube defect accounting for 1-5% of all neural tube defects. Very few cases have been reported in literature. We present a 4day old female patient who presented with a cervical cystic lesion. There was no CSF leakage or sign of infection at presentation. On further evaluation, other congenital abnormalities were excluded by thorough clinical and radiological examination. The patient had normal movement in all four limbs pre-operatively. Intra-operatively, the CSF filled cystic lesion had a hollow fibro-neurovascular stalk extending through the vertebral defect at the level C2-C3. This stalk is thought to be non-functional in most cases in literature but because we could not establish functional status, the hollow stalk was repaired and buried in the surrounding tissue after dura repair. Intra-dural exploration was done to remove adhesions up to the level of the defect to reduce the risk of tethered spinal cord syndrome. Post-operatively, the patient had full power in all limbs with no CSF leak or surgical site infection.

INTRODUCTION

Meningomyelocoele is a neural tube defect with protrusion of the neural tissue and the meninges through a vertebral defect. These can occur anywhere along the spine. The lower meningomyelocoele include lumbosacral and thoracolumbar whereas the high meningomyelocoele are cervical and upper thoracic. The lower meningomyelocoele are more common and maybe associated with neurological deficits. On the other hand, cervical meningomyelocoele are very rare making up 1-5% of all neural tube defects[1]. The embryological cause for meningomyelocoele is failure of closure of neural tube during third week of gestation leading to the constellation of defects. The open neural tube is continuous with the surface of the skin, for this reason infants with meningomyelocoele are at risk of bacterial meningitis and myelitis. Cerebrospinal fluid leak is commonly observed. The major indication for early operative repair within 48hr of delivery is prevention of infection.

Literature review

Cervical myelomeningocele (CMMC) is an extremely rare condition, accounting for 1 to 5% of all neural tube defects[1]. Cervical myelomeningocele differs structurally and clinically from myelomeningocele in distal areas and has a more favourable outcome[1,2]. The authors thought that some trivial neurological deficits in CMMC were caused by the late and limited neurulation abnormality during its development [1]. The management strategies of CMMC are early surgical treatment with standard micro-neurosurgical techniques to prevent the development of neurological defects. It is safe and effective to adopt surgical excision of the lesions with intra-dural exploration of the sac to release any potential adhesion bands [3,2]. Although the cMMC has more favourable outcome, spinal cord dysfunction has to be considered in growing children due to persistent tethering or re-tethering, therefore regular neurologic and urodynamic investigations are of particular importance[1,4].

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The definition and classification of cervical meningomyelocoele has evolved over the years. The earlier terms used to describe CMMC included syringoceles [5] and syringomyeloceles [6]. Maclone *et al.* defined CMMC as closed spinal dysraphism characterized by skin-covered posterior midline mass, a narrow posterior spina bifida and cerebrospinal fluid-filled cyst[7]. Steinbok *et al*. in the early 1900s proposed the hypothesis of limited dorsal myelochisis and divided CMMC as meningoceles and myelocystoceles [8]. Pang *et al.* proposed a classification system based on the internal structure of the cystic lesions, dividing them into limited dorsal myelochisis containing fibro-neurovascular stalk in a dural sac and split cord malformations containing two hemi-cords in a dural sac [9]. Salomao *et al*. divided these into: 1. Cystic spinal dysraphism of the cervical and upper thoracic region with a stalk of neuroglia or fibro-vascular tissue, 2. Myelocystoceles consisting of a second ependymal-lined cyst herniated inside a meningocele and 3. Cystic spinal dysraphism of the cervical and upper thoracic region without a stalk or true meningocele[10]. Habibi *et al.* and Rossi *et al.* classified cervical myelomeningoceles into two sub groups: fibro-neurovascular stalks and myelocystoceles. However, a new multicentre study has defined all lesions as cysticspinal dysraphism of the cervical and upper thoracic region, and divided them into three subgroups: cystic spinal dysraphism of the cervical and upper thoracic region with a stalk of neuroglia or fibro-vascular tissue; myelocystoceles consisting of a second ependymal-lined cyst herniated inside a meningocele; and cystic spinal dysraphism of the cervical and upper thoracic region without a stalk or true meningocele[11].

The occurrence of hydrocephalus with cMMC is reported by several case studies but no standard incidence has been reported as no huge study has been conducted. In a follow-up study of five patients, two (40%) developed hydrocephalus [1]; another report showed that seven (38%)out of 18patients had hydrocephalus [4]. The study of eight patients by Steinbok and Cochrane [8], revealed hydrocephalus in five (56%) patients. In a study of 16 patients, Habib et al, found a 50% occurrence of hydrocephalus in the series and therefore recommended that preoperative brain imaging is performed to assess the evidence of hydrocephalus in any patient who has spinal dysraphism[11].Magnetic resonance imaging is the modality of choice in these lesions, which helps to distinguish the subtypes of CMMC and to identify the associated anomalies of central nervous system like hydrocephalus, split cord malformations, tethering and syringomyelia[12].

The outcome of patients with CMMC is favourable with regards to the neurologic, orthopaedic and urologic problems compared with lower neural tube defects. However, the burden of repeated examinations and therapies is considerable and induces high costs, therefore prevention with peri-conceptual folic acid is a crucial issue in CMMC [1].

DESCRIPTION OF CASE REPORT

The patient was admitted at 4 days post-delivery as a referral from the local hospital for a swelling on the back of the neck. This was the 5th child. The other children had no congenital abnormalities. The mother had attended antenatal care visits late in pregnancy but had never taken folic acid. No obstetric scans had been done prior to delivery.

Examination

Stable neonate with a cystic midline posterior cervical lesion approximately size of a tennis ball (figure 1). It had normal skin covering the lower half

Figure 1: Pre-operative showing lesion gross anatomy with normal skin covering the lower half and purple membrane covering the upper half

and a purplish membrane covering the top half. There was no sign of cerebrospinal fluid leak or infection at the site. There was full range of motion in all limbs.

There were no other obvious congenital abnormalities noted.

Head circumference 37cm

Investigations

CT-Scan cervical spine- Defect at C2-C3 with protrusion of spinal cord and meninges

CT-Scan brain- No hydrocephalus or associated Chiari malformation

Full blood count was normal

Abdominal ultrasound- Normal

Treatment

Pre-operative

1. Antibiotics- Benzyl-penicillin and gentamycin
2. Myelomeningocele care- daily cleaning with iodine and dressing with paraffin gauze

Intra-operative

Meningomyelocoele repair was done in theatre on day3 of admission- The lesion was opened, a fibrous strand of neural tissue extending from the defect and fanning out into the skin covering was found (figure 2). Dura was opened and neural tissue separated from the lesion. Dura exploration was done to remove adhesions down to a level slightly below the defect. Dura was repaired and skin refashioned (figure 3).

Figure 2: Intra-operative showing fibro-neurovascular stalk with dura covering and cystic lesion

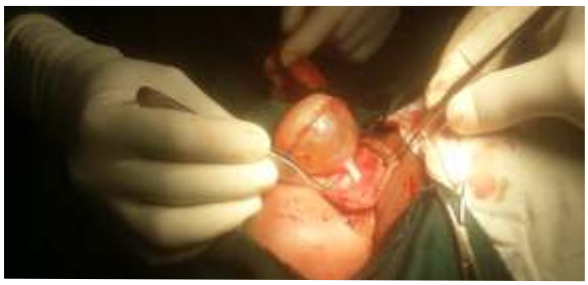

Figure 3: Post-operative, showing the repaired defect

Post-operative

1. Antibiotics- cefotaxime and cloxacillin
2. The patient had good recovery with no surgical site infection or CSF leak. She had good power and movement in all limbs.
3. Discharged after 6days post operative

DISCUSSION

Cervical meningomyelocoele is a rare neural tube defect with an incidence of 1-5%. Our patient presented at 4 days of life. The lesion was covered with normal skin on the lower half and a pinkish membrane on the top half. From the literature reviewed most of the cases had a similar gross appearance. There was no CSF leakage at the time of presentation or any sign of infection. This gave us a bit of time to investigate fully and exclude other congenital abnormalities. However, the presence of a leakage or infection is an indication for urgent repair. The repair was done on day 3 of admission. The intra-operative findings where that of a fibro-neurovascular stalk protruding the vertebral defect. Some literature suggests that this fibro-neurovascular stalk is non-functional and can be safely excised but it was not possible to assess the function of the stalk. Hence in this case the stalk was dissected from the wall of the meningomyelocoele and hollow defect on the stalk closed. The stalk was freed and buried in the surrounding tissues.

The spinal canal at the defect was explored to remove adhesions that may cause tethering with elongation of vertebral column. At the time of operation, the dura was repaired and no CSF leak was noted. As noted in most cases in literature, there was adequate skin cover to repair the defect. Post operatively, the neonate had movement in all 4 limbs. However, this does not completely exclude the possibility of neurological deficits later in life as re-tethering can occur. Hence the need for long term follow-up in these patients.

Folate deficiency is a major risk factor for neural tube defects. In this case, the patients mother had not taken folic acid supplements during this pregnancy. In her other 4 pregnancies prior, she had folate and ferrous supplementation with normal outcome.

CONCLUSION

Due to it's a rare occurrence, literature is scanty on the subject of cervical meningomyelocoele and hence the need for a more vigilant case reporting and patient follow up. Cervical meningomyelocoele has a better neurological outcome compared to lower lesions. However, neurological deterioration may occur later in life due to cord tethering. In the presence of CSF leakage or signs of infection, the lesion must be repaired urgently. Excision of the lesion with intra-dural exploration of the sac to release any potential adhesion bands, is the recommended management. In addition, identification and management of associated congenital abnormalities is invaluable as it affects prognosis.

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