Case Report

Successful intervention of meningomyelocele associated with type 2 Arnold Chiari malformation and hydrocephalus – A case report

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ABSTRACT

Myelomeningocele, also known as open spina bifida, is characterized by non-closure of neural tube during embryonic development causing herniation of meninges and spinal cord through the vertebral defect. The risk of recurrence after one affected child is 3–4% and increases to 10% after two affected children. Meningomyelocele may rarely also be associated with Arnold Chiari malformations and hydrocephalus. However, most cases of Arnold Chiari malformation may remain asymptomatic.

Key words: Arnold Chiari malformation, Hydrocephalus, Meningomyelocele, Spina bifida

Myelomeningocele is known as open spina bifida and is associated with significant morbidity and mortality [1]. It is characterized by non-closure of neural tube during embryonic development causing herniation of meninges and spinal cord through the vertebral defect [1]. It is more commonly seen in females more than males. The risk of recurrence of neural tube defect (NTD) after one affected child is 3–4% and increases to 10% after two affected children [2]. The prevalence of NTDs in India is 4.5/1000 live births [3]. Folate is essential for the prevention of NTDs. Maternal periconceptional folate supplementation reduces the incidence of NTDs [4]. Meningomyelocele may rarely be associated with Arnold Chiari malformations and is of four types, classified according to the level of descent of the tonsillar herniation [5].

CASE REPORT

A 4-month-old male infant presented to the pediatric clinic with swelling at the lower back since birth. The swelling gradually increased in size and parents noticed skin excoriation and leaking of clear fluid from superior aspect of the swelling for the past 1 month and fever for 10 days. Parents also noticed reduced right lower limb movements. There was no history of consanguinity. The patient had no history of convulsions or focal neurodeficit or cranial nerve involvement. The mother was not antenatally registered and did not undergo any antenatal screening for congenital defects nor did she consume iron and folic acid supplements. The child was born at full term through spontaneous vaginal delivery, cried immediately at birth and weighed 3 kg, birth events were not significant.

On examination, the child's vitals were stable, head-to-toe examination revealed macrocephaly, anterior fontanelle measuring 3 cm × 3 cm, posterior fontanelle was closed, a single café au lait sot was noted on the right scapular region measuring 3 cm × 4 cm. The swelling located in lumbosacral region was noted to be 10 cm × 8 cm in size, with skin excoriation on superior aspect of swelling, and leakage of cerebrospinal fluid from the defect (Fig. 1). Central nervous system examination revealed reduced tone and power in the right lower limb, and normal deep tendon reflexes. Detailed ophthalmic examination revealed bilateral sunset sign and Collier's sign. Fundus examination showed Grade I papilledema.

Routine blood investigations were within normal limits. Swab from site of cerebrospinal fluid leak was positive for *Methicillin-resistant Staphylococcus aureus*. Magnetic resonance imaging (MRI) brain showed dilated lateral and third ventricles, findings suggestive of benign enlargement of subarachnoid space in infancy, and tonsillar descent of 11.3 mm with crowding neural structures at the level of foramen magnum, which was suggestive of Arnold Chiari malformation Type 2 (Fig. 2). MRI spine findings were suggestive of lower lumbar myelomeningocele with defects in posterior elements of lower lumbosacral spine (Fig. 3).

He was started on intravenous antibiotics meropenem and vancomycin. Surgical repair of meningomyelocele defect was

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done urgently with detethering of spinal cord with dural repair under general anesthesia. Postoperatively, the patient developed hypertension and persistent vomiting. Due to raised intracranial pressure, computed tomography brain was done on POD 2, which revealed hydrocephalus and tonsillar descent of 5 mm that had reduced from 11.3 mm than previous MRI (Fig. 4). The patient was started on oral acetazolamide in view of hydrocephalus with hypertension.

After completion of antibiotic course, the patient was discharged and initially followed up twice weekly and then once a month. No motor deficit was present postoperatively. Physiotherapy was started for the patient, after which gradual improvement in tone and power was noted.

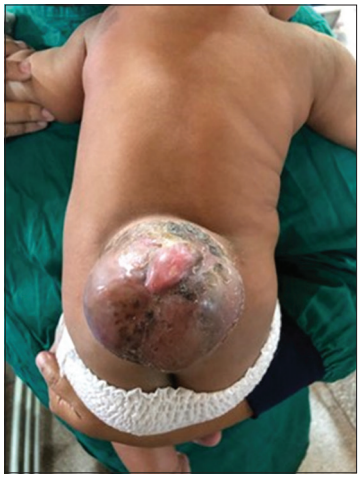

Figure 1: Clinical picture showing meningomyelocele as described above with skin excoriation and cerebrospinal fluid leak

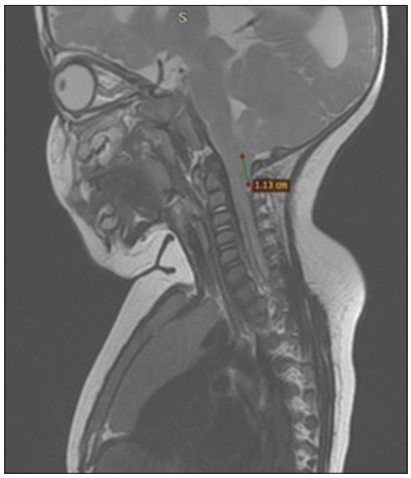

Figure 2: Magnetic resonance imaging spine image showing tonsillar descent of 11.3 mm suggestive of Arnold Chiari malformation Type 2

DISCUSSION

There are various types of NTDs. Incomplete fusion at the caudal end of neural tube leads to the formation of meningomyelocele around the 26th day of gestation [1]. Folate has an essential role in the prevention of NTDs. The incidence of NTDs can be reduced through folate supplementation in the periconception period [6]. Multiple risk factors have been linked to the defect, some of which are parent or sibling with NTD, trisomies 18 and 13, Meckel–Gruber syndrome, HARD syndrome, VACTERAL and VATER associations, and some maternal factors such as drug intake, alcohol abuse, and maternal nutritional deficiencies [1].

Meningomyelocele may also be associated with Arnold Chiari malformations type 2 and 4. Type 1: This is the most common type, where tonsillar herniation is up to 5 mm through the foramen magnum, often associated with syringomyelia. Type 2: Also known as classic Chiari malformation and is less common and characterized by the descent of cerebellar tonsils and vermis with the brain stem and fourth ventricle into cervical spinal cord. It is associated with meningomyelocele and hydrocephalus. Patients may become symptomatic in early infancy. Type 3: It is the most serious type, having occipital or cervical encephalocele and a wide foramen magnum. Type 4: It rarely occurs and has cerebellar hypoplasia or aplasia and tentorial hypoplasia. Our patient had Type 2 Arnold Chiari malformation with hydrocephalus with tethered cord, which is rare as reported in literature [7].

Surgical repair of defect with detethering of cord done shortly after birth is the treatment for meningomyelocele. Postoperatively, the patient should be started on physiotherapy and routinely

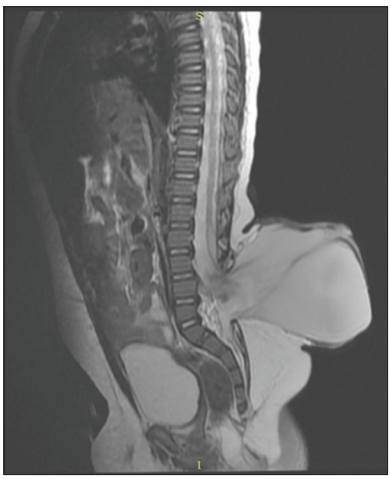

Figure 3: Magnetic resonance imaging spine image showing meningomyelocele with tethered spinal cord

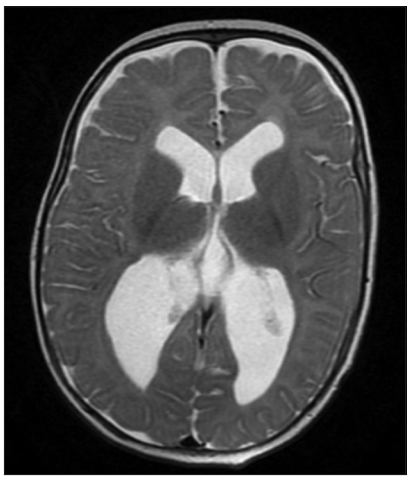

Figure 4: Magnetic resonance imaging brain image showing hydrocephalus

followed up for monitoring of development. Our patient underwent surgical repair of meningomyelocele with detethering of cord at 4 months of age. The surgery was delayed due to severe acute respiratory syndrome coronavirus 2019 pandemic situation, after which he was started on physiotherapy.

Mortality rate for children born with meningomyelocele is 1% per year between 5 and 30 years [1]. Life-threatening complications may occur at any age. Motor deficits, bladder and bowel dysfunction, learning problems, and seizure disorder are more common in survivors than general population. Majority may have normal intelligence. Periodic multidisciplinary follow-ups are recommended.

It is recommended that all women of reproductive age should consume 0.4 mg of folic acid daily which is essential for reducing the risk of NTDs [6]. Ideally, folic acid supplementation should be initiated at least 1 month before conception and continued till the 12th week of gestation, when neurulation is complete. The supplementation in women with one previously affected offspring is 4 mg/day, starting 1 month before planned conception [4]. Ensuring Vitamin B12 sufficiency is important while undertaking folate supplementation [4]. Foods that are rich in folic acid should be consumed. Fortification of foods and cereals with folic acid can also help with daily supplementation. Certain drugs that cause meningomyelocele should be avoided during pregnancy, such as valproic acid, phenytoin, and carbamazepine.

CONCLUSION

Meningomyelocele is an open NTD which can be detected early by regular antenatal screening, and therefore, early intervention should be undertaken to reduce complications and improve the prognosis of the infant. Dietary diversity, food fortification, and periconceptional supplementation are recommended for prevention.

REFERENCES

1. Sahni M, Alsaleem M, Ohri A. Meningomyelocele. In: Stat Pearls. Treasure Island, FL: Stat Pearls Publishing; 2021.
2. American College of Obstetricians and Gynecologists. Neural Tube Defects, ACOG Practice Bulletin No. 44. Washington, DC: ACOG; 2008.
3. Allagh KP, Shamanna BR, Murthy GV, Ness AR, Doyle P, Neogi SB, *et al*. Birth prevalence of neural tube defects and orofacial clefts in India: A systematic review and meta-analysis. PLoS One 2015;10:e0118961.
4. Ganesh D, Sagayaraj BM, Barua RK, Sharma N, Ranga U. Arnold Chiari malformation with spina bifida: A lost opportunity of folic acid supplementation. J Clin Diagn Res 2014;8:OD01-3.
5. Hassan A, Yaseen S, Rashid M, Afza R, Kaur M, Javid M. Arnold-chiari malformation: Anatomical variations and latest embryological perspective. Review of literature. Int J Contemp Med Res 2016;3:1489.
6. Chitayat D, Matsui D, Amitai Y, Kennedy D, Vohra S, Rieder M, *et al*. Folic acid supplementation for pregnant women and those planning pregnancy: 2015 update. J Clin Pharmacol 2016;56:170-5.
7. Khoshhal K, Abos M, Waleed E, Essam S, Mustafa AM. Tethered cord syndrome: A study of 35 patients. J Taibah Univ Med Sci 2012;7:23-8.

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