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An unusual case of cerebellar herniation in a neonate secondary to communicating hydrocephalus

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ABSTRACT

The incidence of sepsis causing meningitis is a common finding seen in neonates. Meningitis, if not diagnosed early and intervened on time with adequate amount and duration of antibiotics can lead to catastrophic complications in the neonate, mainly leading to a communication hydrocephalus with mantle thinning, further leading to severe neuro morbidity. This case reports a neonate presenting on the 5th day of life with repeated episodes of vomiting but was treated symptomatically. The patient was further referred to us with incessant vomiting and depressed activity on day of life 24. The patient further had failure to thrive with significant weight loss since birth. General physical examination and routine examination revealed a case of communicating hydrocephalus secondary to sepsis causing meningitis which was left untreated. On further evaluation, neuroimaging revealed cerebellar tonsillar herniation. It is a common phenomenon to expect raised intracranial (ICP) leading to herniation in patients with a closed fontanelle. Here, we report a case of gross, communicating hydrocephalus causing cerebellar herniation in a paraneonate, which is not a well-known phenomenon in literature.

Keywords: Neonate, Partially Treated Meningitis, Hydrocephalus, Cerebellar Herniation, Subgaleal Shunt, Ventricular dilatation.

1. INTRODUCTION

The rate of neonatal sepsis in India is considered to be one of the highest in the world contributing to 5.7% to neonatal mortality (NHM). Bacterial sepsis and meningitis continue to be leading causes of morbidity and mortality in neonates. Neonates that survive sepsis may have long term neurological sequelae as a result of central nervous system (CNS) infections, white matter damage secondary to systemic inflammation and hypoxic damage due to septic shock, persistent pulmonary hypotension or chronic lung disease (Cloherty). Neonatal meningitis can be classified into early onset meningitis

(EOM) or late onset meningitis depending upon the time of presentation with EOM presenting in the first week for like and LOM presenting anytime in the last three weeks of neonatal period (Khalessi and Afsharkas, 2014). Prevalence of LOM is more than EOM. Any neonate suspected to have sepsis should undergo a lumbar puncture and should be evaluated for meningitis. Meningitis, if not treated on time and for designated amount with injectable antibiotics, leads to severe consequences like hydrocephalus, brain abscess, infarction and subdural effusion leading to long-term sequelae like developmental delay, cerebral palsy, late onset seizures, hearing loss, cortical blindness and cognitive impairment. Untreated or partially treated meningitis leading to communicating hydrocephalus is common sequelae.

As a basic knowledge, any CNS pathology in neonates and infants, because of an open anterior fontanelle (AF) and sutures, does not lead to a rise in ICP as the increasing pressures are compensated by an increase in head size. This is clinically seen as a bulging AF or opening of the sutures and in patients with closed fontanelle, it presents with signs of headache, vomiting, blurring of vision and in severe cases leads to coma and sudden death, the most common cause being subfalcine herniation also known as a midline shift (Munakomi and Das, 2022). In an acute setting when there is overwhelming elevation in intracranial pressures, the neonatal skull fails to compensate leading to signs and symptoms or raised ICP. This phenomenon is rarely seen in this particular age group and has been reported secondary to only congenital intracranial tumors or intracranial bleeds. However, neonatal meningitis leading to gross hydrocephalus causing raised ICP and cerebellar herniation is a rare, unreported phenomenon but its existence is certain through this particular case report needing further research and study.

2. CASE REPORT

Our patient, a female child, weighing 3 kgs at birth was born to a primigravida mother at 39 weeks of gestational age via normal vaginal delivery in a hospital setup in rural India. Baby cried immediately after birth, receiving essential newborn care. Patient was discharged with mother on day of life-3. On the 15th day of life, the patient developed 3-4 episodes of vomiting of milk associated with irritability. The patient was taken to a local doctor, a week later, in view of persistent vomiting and excessive irritability. Patient was treated symptomatically with prokinetics giving symptomatic relief. Another fortnight later, the patient was again taken to a tertiary center, now with complaints of recurrent episodes of vomiting for 2 days, not responding to oral medications and decreased activity. The patient was also reported to have weight loss. Ultrasonography (USG) abdomen revealed no obvious abnormality. Neurosonogram done was reported as mild hydrocephalus with bilateral cortical thinning. The patient was further referred to us for further management. On admission, the patient weighed 2.2 kgs, height of 51cm and had a head circumference of 36.5 cm which was just above the 50th percentile (36.4cm) expected at 4 weeks of age as per WHO growth chart for girls (Figure 1).

Figure 1 Clinical image of the neonate showing evidence of increased head circumference with dilated veins and having scissoring of lower limbs

Patient's Heart rate was 150 beats per minute with a respiratory rate of 44 cycles per minute, peripheral pulses were well felt with good peripheral circulation and a normal capillary refill time. Systemic examination of the nervous system revealed bilaterally reactive pupils; widely open, bulging anterior fontanelle with hypertonia of bilateral lower limbs and exaggerated deep tendon reflexes. Other systemic examinations revealed no significant abnormality. Routine blood investigations done showed Hb-13.7 gm/dL, TLC - 16,900 cells/cumm, platelets- 7.1 lakhs/cu.mm, C-reactive protein was negative, renal and liver function tests were within normal limits. Serum electrolytes were also normal with serum sodium - 133 mmol/L, serum potassium-3.9 mmol/L and serum calcium-9.3 mg/dL. Serum Ammonia and Lactate don to rule out Inborn Errors of Metabolism (IEM) was also normal. Lumbar puncture was done which revealed 35 cells/cu.mm, predominantly polymorphs (60%). CSF proteins were grossly elevated to 4200, CSF glucose of 39 mg/dL against a parallel blood sugar of 156 mg/dL and LDH of 200 IU/L. Patient was started on injectable antibiotics in meningitis dosage. Neurosonogram repeated was suggestive of gross dilatation of bilateral lateral ventricle, third ventricle and fourth ventricle with thinning of brain parenchyma suggestive of communicating hydrocephalus. Patient further developed convulsive episodes, not responding to first line anti-epileptics. Antibiotics were also upgraded but the patient was eventually intubated in view of status epilepticus and was started on Inj. Midazolam infusion, bringing seizure control. Patient was slowly weaned off anti-epileptics and was extubated over a course of 6 days. In the meantime, the patient continued to have increased head circumference at a rate of 0.5 over 1 week. Patient was started on oral Diamox. Neurosurgery opinion was sought and the patient went through a therapeutic as well as diagnostic ventricular tap (Figure 2).

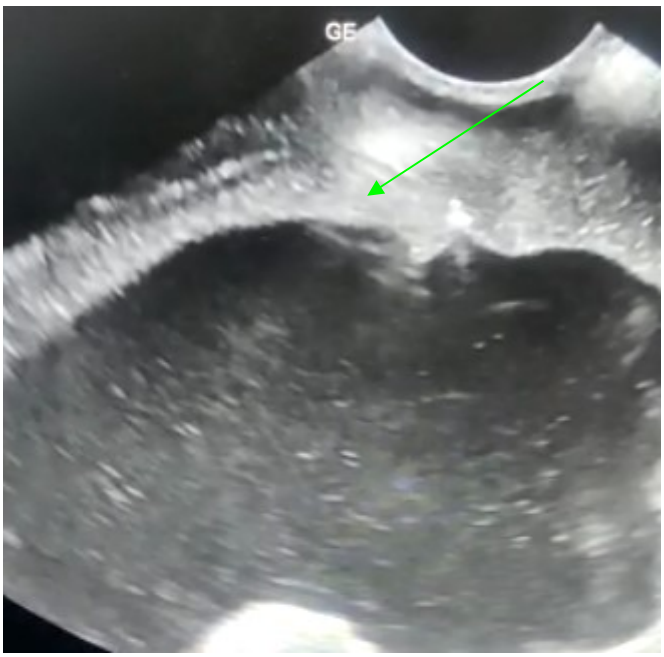

Figure 2 Neurosonogram showing grossly dilated lateral ventricle while undergoing ventricular tapping (green arrow)

CSF samples from the ventricles taken showed 300 cells/cu.mm with 60% lymphocytic predominance, CSF proteins of 1800, CSF glucose of 43 mg/dL. CSF on culture showed growth of *Klebsiella pneumoniae* with more than 10^5 CFu/ml. Antibiotics were administered and planned for a minimum 21 days according to the sensitivity report. The patient, in the due course, showed signs of raised ICP in the form of hypertension with BP recordings of 100/60 mmHg which lies in between 75th to 90th centile for the given age of 6 weeks in a female which was followed by bradycardia with a baseline, sleeping heart rate of 60-70 beats per minute.

Patient was also started on Inj. Furosemide for hypertension and hydrocephalus. The patient required repeated therapeutic ventricular taps to reduce the intraventricular pressure, which on routine examination showed reducing cell counts and improving CSF proteins with the latest report showing 1-2 cells/cu.mm, CSF proteins of 356 and CSF glucose of 62 against a blood sugar of 132 mg/dL. MRI brain with contrast enhancement done was suggestive of gross dilatation of bilateral lateral, third and fourth ventricles, foramen of monro, bilateral foramen of luschka (Figure 3, 4), cerebral aqueduct with periventricular ooze and thinning of corpus callosum with evidence of few cystic spaces in the right frontal lobe anterior to the frontal horn of the right lateral ventricle due to periventricular leukomalacia (Figure 5) with secondary herniation of the cerebellar tonsil through the foramen of magnum by 6 cm, secondary to meningoencephalitis (Figure 6).

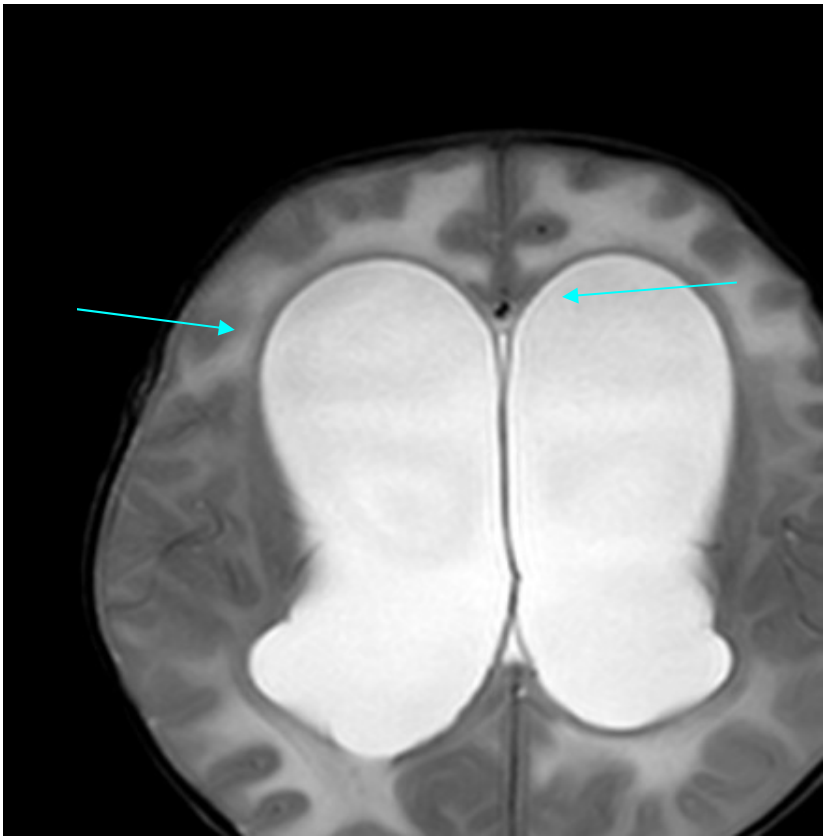

Figure 3 MRI brain contrast enhanced T2 weighted axial view showing gross dilatation of bilateral lateral ventricles

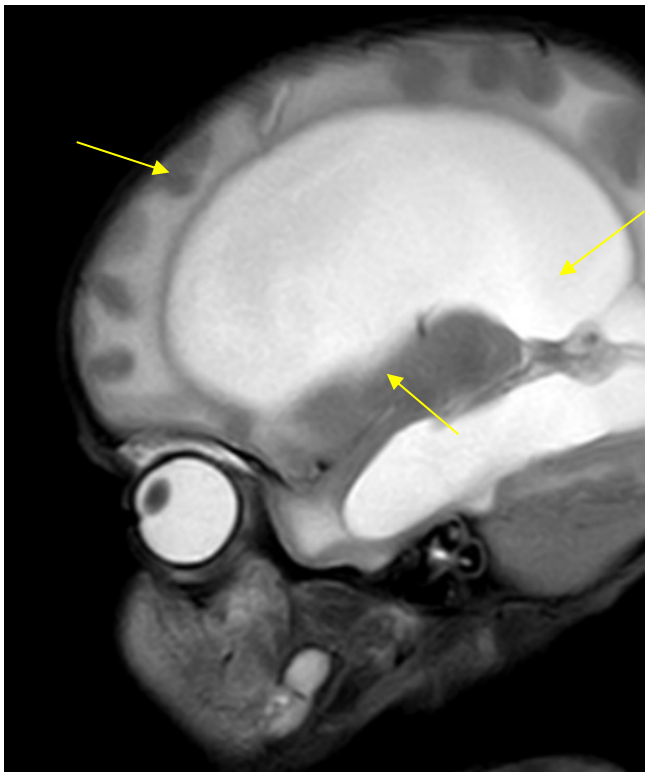

Figure 4 MRI brain contrast enhanced Sagittal T2 weighted image showing gross dilatation of lateral ventricle, foramen of Monro and third ventricle (yellow arrow)

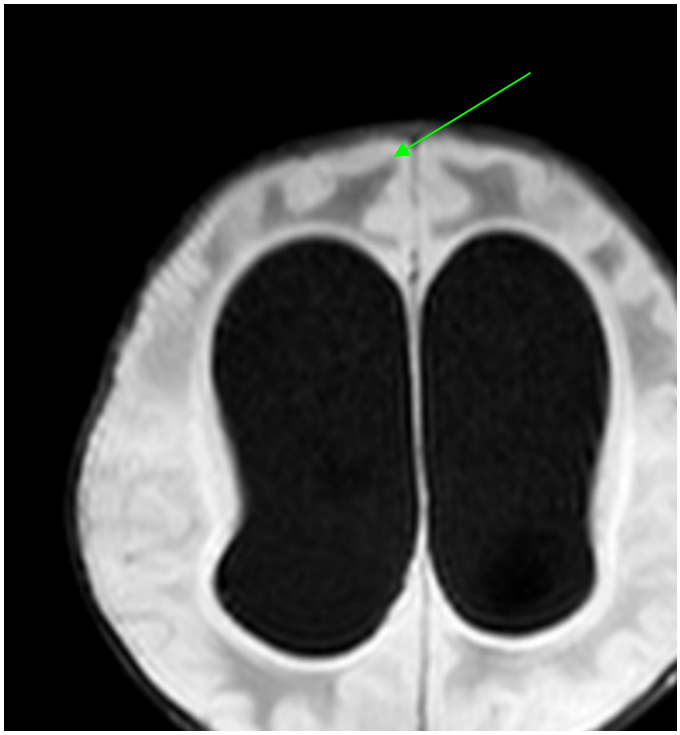

Figure 5 MRI brain contrast enhanced Axial FLAIR sequence showing cystic spaces in the frontal lobe suggestive of periventricular leukomalacia

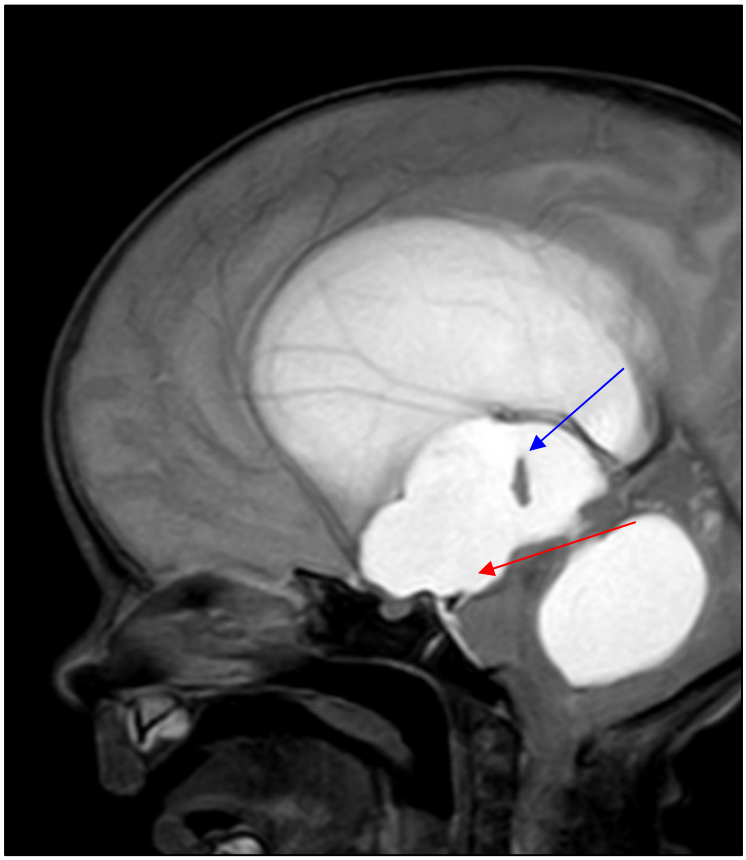

Figure 6 MRI brain contrast enhanced Sagittal T2 weighted image showing dilatation of the fourth ventricle (blue arrow) with cerebellar tonsillar herniation through the foramen magnum (red arrow)

Patient was managed on injectable antibiotics given for 28 days with complete clearance of cells on CSF examination. Neurologist opinion was taken and the patient was advised for subgaleal shunt. Patient was started on neuro physiotherapy in

view of increased tone in bilateral lower limbs. Otoacoustic Emission showed abnormality in both ears, Brainstem Evoked Response Audiometry (BERA) is planned at 3 months of age. Regular weight monitoring was done and milk fortification was added for adequate weight gain. As the CSF proteins significantly reduced, the patient has been posted for a subgaleal shunt placement.

3. DISCUSSION

Hydrocephalus is a disorder in which there is an excessive amount of cerebrospinal fluid (CSF) accumulation within the cerebral ventricles and/or subarachnoid spaces, leading to ventricular dilation and elevated ICP. It is a fairly common problem with complex pathogenesis. In accordance with the Monro-Kellie hypothesis, intracranial matter is made up of 3 components-the blood, the CSF and the brain parenchyma. Considering the skull to be a close, rigid box; a loss or gain in the volume of any of the components occurs at the cost of the other two. ICP in neonates and infants is characteristically lower than seen in adults. The transition of this pressure to equate with adult values takes place around 6-8 years. Neonates, in the initial days are known to lose body weight and water in the first 7-10 days of life. There is associated loss of volume and turgor of the brain, but the level at which this happens is uncertain. Thus, as a physiological adaptation, there is a physiological hydrocephalus seen in neonates compensating the loss in brain volume (Welch, 1980).

As a buffer system, in neonates and even in infants, till the AF closes physiologically between the ages of 9 months to 18 months, elevated intracranial pressure is dissipated due to an open fontanelle and sutures. Citing this, adverse events and sequelae secondary to raised ICP are not expected or commonly seen in neonates. Post-infectious hydrocephalus is known to be one of the most common sequelae to partially treated CNS infections seen early in the course of disease. Hydrocephalus secondary to CNS infection can be perinatally acquired or postnatally acquired. Perinatally infected neonates presenting with hydrocephalus are seen in congenital toxoplasmosis or congenital CMV infection. Meningitis caused by gram negative bacteria is more likely to lead to multiloculated hydrocephalus as they cause ventriculitis (Garg and Gupta, 2021).

Postnatally, sepsis and meningitis leading to hydrocephalus in neonates is most commonly caused by *Escherichia coli* and group *B streptococci* resulting from maternal tract infection. In neonates with late-onset sepsis, meningitis is the main feature and is mainly seen in preterm neonates with group *B streptococci* and *staphylococci* being the causative organisms (Chatterjee and Chatterjee, 2011). In these cases, the source may be maternal or nosocomial. Most cases of meningitis causing hydrocephalus in the neonatal period present with signs of depressed activity and refusal to feed; vomiting, fever, dehydration and seizures associated with increasing head circumference, if noticed earlier, with a bulging AF which is wide open, to name a few. Raised ICP in neonates is a rare phenomenon. A late, but classical sign of raised ICP is the Cushing's reflex characterized by Cushing's triad of a wide pulse-pressure hypertension followed by bradycardia and altered respiratory pattern. This is usually seen late in the disease and carries a poor prognosis for the patient (Dinallo and Waseem, 2022). The most deleterious effect of raised ICP in cerebral herniation.

Compression or traction on neural and vascular structures can lead to injury from herniation of brain tissue. Herniation can happen in four locations in the cranial cavity as a result of a pressure difference between the intracranial compartments namely transtentorial herniation, subfalcine herniation, foramen magnum and posterior displacement of frontal lobes over the lesser wing of sphenoid. As discussed before, the open anterior fontanelle in neonates and infants acts as a buffer and the incidence of raised ICP being reported in this population is few. One such case reported by Wong et al., (2010) mentioned a neonate diagnosed antenatally with intracranial tumor. Postnatally, the patient showed signs of raised ICP with bulging fontanelle with hydrocephalus and Cushing's triad. The neonate succumbed on the operating table to massive hemorrhage during tumor resection.

Histopathologically, the tumor was reported as an intracranial atypical teratoid/rhabdoid tumor (Wong et al., 2010). In another case reported by Khansare et al., (2015) mentions a term neonate with intrauterine growth restriction presenting with seizures poorly controlled with anti-epileptic drugs. The neonate soon showed signs of raised ICP like bradycardia, altered respiratory pattern, anisocoria with non-reactive pupils and decorticate posturing. The urine routine was positive for CMV infection and MRI brain was suggestive of large hemorrhage in the left frontal lobe secondary to thrombocytopenia with a midline shift of 5mm and suture dehiscence. This case clinically represented the possibility of uncal herniation (Khansare et al., 2015). Apart from tumor and intracranial bleeds causing a raised ICP and herniation, patients suffering from perinatal asphyxia and having hypoxic ischaemic encephalopathy have also been demonstrated to have a similar course.

Soliman et al., (2019) reports a case of a term baby delivered through vacuum-assisted normal vaginal delivery with meconium-stained liquor and delayed cry having a poor Apgar score requiring intubation with cord blood gas suggestive of severe birth asphyxia. Initially presenting with subtle seizures but eventually showing signs of shock and further deterioration with absence of spontaneous movement, hypotonia and anisocoria. There was increase in the head circumference from birth with a bulging anterior fontanelle followed by bradycardia. CT brain done showed features of uncal and tonsillar herniation with the neonate succumbing

over the next 2 hours (Soliman et al., 2019). There is also a case reported of Cleidocranial Dysplasia in a neonate having asymptomatic tonsillar herniation where the patient initially presented with urosepsis at 24 hours of life but then progressively developed apneic episodes and neuroimaging of the brain showed posterior fossa crowding with tonsillar and uncal herniation (Myers et al., 2014). Though rare, cases of various parts of brain herniation have been documented in literature citing various etiologies. However, no case has been reported till date suggesting raised ICP leading to cerebellar herniation secondary to only post-infectious hydrocephalus, one of the most commonly encountered conditions in neonatal practice. The only plausible pathophysiology behind this could be dilated ventricles causing a mass effect over the brain parenchyma leading to herniation. This raises an alarm towards suspecting and timely and aggressively treating patients suspected or diagnosed with meningitis to prevent such deleterious effects.

4. CONCLUSION

Meningitis, its incidence presenting symptoms and complications, though commonly known, is often underestimated. Any neonate presenting with sepsis should undergo a lumbar puncture to rule out meningitis. A multidisciplinary approach helps in better patient care and pediatric neurologist opinion should be taken early in the course for a better outcome. Keeping in mind with the already known consequences of missing or undertreating this condition with the new added information provided by this case report which clearly signifies the ability of a gross hydrocephalus to cause elevated ICP leading to herniation, it is generally advisable to rule out or provide timely treatment to neonates having sepsis and suspected to have meningitis to avoid serious long-term morbidity and mortality.

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Author contribution

Dr Iyer Lavanya Ramakrishnan - Drafting of the manuscript, data analysis and edition.
Dr Mahaveer Lakra - Idea behind the article and final manuscript edition and approval.
Dr Bhavana Lakhkar - Expert opinion on the subject.
Dr Nishant Raj - Acquisition of appropriate radiological images.
Dr Shikha Kakkat - Data acquisition and involvement in drafting part of the manuscript.
Dr Anirudh Komareddy - Acquisition and edition of clinical pictures.

Consent

Well informed, written consent from the child's parents was obtained.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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