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Case Report

Congenital Basal Meningocele: An Unusual Cause of Nasal Obstruction in Early Life

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Abstract—Basal meningoceles are rare congenital defects that can cause nasal obstruction and often clinically occult until they result in life-threatening complications. Knowing the clues to early diagnosis, management, and complications is essential. Case: A 7-day-old baby girl was referred to our hospital because of high fever and dyspnoea, and the baby was diagnosed with pneumonia, lip tie, cup ears and suspicion of laryngomalacia. The patient got dyspnoea with stridor when drinking, and it decreased when her mouth was open. The suction catheter could not enter through the left choana. The nasal endoscopy showed an elevation of the hard palate. A 3-dimensional facial CT scan demonstrated a transsellar–transsphenoidal meningocele protruding into the left nasal cavity. A diluted liquid came out from the left nose with a yellowish-clear colour, and the baby showed a high-pitched cry. Bacterial meningitis was established from cerebrospinal liquor analyses. After meningitis treatment, surgical repair to meningocele reposition and bone defect repair was done at 40 days. Conclusion: In our case, the nasal obstruction was not detected from the beginning of birth, and it led to delays in finding the cause. Basal meningocele in this case was accidentally diagnosed by a facial CT scan exploring the cause of choana atresia. It's essential to detect choana atresia since birth, explore the etiology immediately, and manage it well to prevent life-threatening complications.

Keywords: *nasal obstruction, congenital basal meningocele*

Abstrak—Meningocele basal merupakan kelainan kongenital langka yang dapat menyebabkan obstruksi hidung yang secara klinis sering tersembunyi sehingga baru diketahui saat sudah terjadi komplikasi. Oleh karena itu sangat penting untuk mengetahui cara menegakkan diagnosis dini agar dapat diberi tata laksana yang tepat untuk mencegah terjadinya komplikasi yang mengancam nyawa.-Kasus: Seorang bayi perempuan berusia 7 hari dirujuk ke rumah sakit kami karena demam tinggi dan dispnoea dan bayi itu didiagnosis sebagai pneumonia, ikatan bibir, telinga cangkir dan kecurigaan laringomalesia. Pasien mengalami dispnea dengan stridor saat minum; dan menurun ketika mulutnya terbuka. Kateter hisap tidak bisa masuk melalui choana kiri. Endoskopi hidung menunjukkan peningkatan langit-langit keras. CT scan wajah 3 dimensi menunjukkan transsellar – meningocele transsphenoidal yang menonjol ke dalam rongga hidung kiri. Cairan encer keluar dari hidung kiri dengan warna bening kekuningan, dan bayi itu menunjukkan tangisan nada tinggi. Meningitis bakteri ditetapkan dari analisis cairan serebrospinal. Setelah meningitis diobati, perbaikan bedah reposisi meningocele dan perbaikan cacat tulang dilakukan pada usia 40 hari. Kesimpulan: Dalam kasus kami, meningocele basal secara tidak sengaja didiagnosis dengan CT scan wajah yang mengeksplorasi penyebab choana atresia. Sangat penting untuk mendeteksi choana atresia sejak lahir, segera mengeksplorasi etiologinya, dan mengelolanya dengan baik untuk mencegah komplikasi yang mengancam jiwa.

Kata kunci: *obstruksi hidung, meningocele basal kongenital*

INTRODUCTION

Basal meningoencephalocele is a rare congenital anomaly characterized by the herniation of brain tissue through the skull, with an incidence of 1 per 40,000 live births [1]. Southeast Asia is an endemic focus with an incidence of 1 per 6000 live births [2]. Compared with other encephaloceles, meningoceles contain meninges only and are classified according to the location of the bony defect: trans ethmoidal, sphenoethmoidal, transsphenoidal, spheno-orbital, and sphenomaxillary [3]. Basal meningoceles present as masses in relation to the nose and may cause external nasal deformity and hypertelorism. Congenital basal meningoceles are often clinically occult; however, a diagnosis makes it possible to perform the necessary surgical repair or prevent fatal episodes of meningitis [4,5].

A case of congenital basal meningocele in a neonatal-age baby girl will be reported here. This case report provides a lesson on the importance of early detection of obstruction in the upper airway especially in the nose in every newborn, and investigates the cause of the obstruction. Finding the etiology is important so that proper management can be done and its life-threatening complication can be prevented. The case report also describes the management, outcomes and progress up to 2 years later.

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CASE

A 7-day-old baby girl was referred to our hospital because of high fever and dyspnoea. From history, she had a fever, cough and rhinorrhoea for 3 days. Physical examination showed nasal flare, stridor, chest retraction and rhonchi in both her lung fields. Chest X-ray showed infiltrate in both sides of the lungs. The baby also had hypertelorism, upper lip tie and cup ears. Her mouth was always slightly open, especially on the upper side (Figure 1). The dyspnoea and stridor were getting worse when she drank with the bottle. There was difficulty inserting a suction catheter into both sides of her nose. From birth history, the baby was delivered by caesarean section in another hospital. There was no information about nasal obstruction after birth, and her parents had never known about this problem. The baby was diagnosed with pneumonia, suspicion of laryngomalacia and choana atresia, Upper Lip Tie and Ears Cup.

Figure 1. Baby's face with cerebrospinal fluid rhinorrhoea and lip tie.

The pneumonia was managed by antibiotics and supportive therapy such as hydration and bronchial toilet. The patient was consulted to an Ear, Nose and Throat Specialist to explore the upper respiratory obstruction. She was consulted to a Plastic Surgery Specialist because of her lip tie and ear cup.

In a few days, the pneumonia improved, but patients still seemed to have breathing difficulty with stridor and a slight open mouth condition. Based on rhinoscopy, the baby was suspected of having choana atresia. A nasal endoscopic showed elevation of the hard palate, closing the choana, resulting in total right choana atresia and partial left choana atresia. Then, a 3-dimensional facial CT scan demonstrated a transsellar–transsphenoidal meningocele protruding into the left nasal cavity with basal anterior skull defect $\pm$ 0.78 cm width and $\pm$ 2.16 cm length (Figure 2).

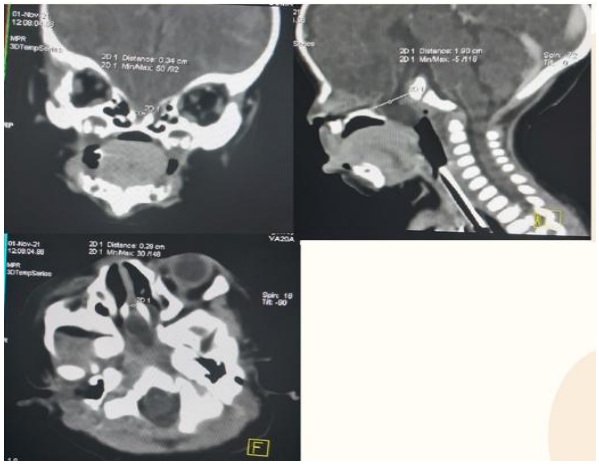

Figure 2. Head CT: Transsellar – transsphenoidal meningocele with anterior base skull defect.

During hospitalization, the patient still had a fever and high-pitched cry, and a diluted liquid came out of the left nose with a yellowish-clear colour several times (Figure 1). Then, a Lumbar puncture was carried out, and signs of bacterial meningitis were obtained from the analysis of cerebrospinal liquor. The patient was treated for meningitis for up to 21 days. Base skull defect closure with a surgical approach was successfully done at the age of 40 days by open craniotomy. The baby could have already breathed better, and there was no complication.

Five months after surgery, the baby developed a prolonged cold cough, then developed a high fever and decreased consciousness. A diluted liquid came out from her left nose again. MRI of the head was done and showed a leak of cerebrospinal fluid around the titanium, and the baby had meningoencephalitis. Leak closure was performed by a neurosurgeon then. Two months later, the baby was inserted with a ventriculoperitoneal (VP) shunt because the baby had hydrocephalus due to blockade in the brain's ventricles. Postoperatively, the baby has repeated seizures but can be overcome with anti-seizure drugs.

Currently, the patient can do daily activities well by staying attached to VP-shunt and taking anti-epilepsy drugs with normal growth and development.

DISCUSSION

Nasal obstruction encompasses anything that hinders the airflow in and out of the nose, affecting one or both nasal passages. Nasal obstruction is usually caused by either swelling of the nasal tissue or an anatomical blockage, which results in a narrowing of the nasal cavity and the feeling of the passages being congested. Nasal obstruction can lead to sleeping disorders, snoring, and obstructive sleep apnoea (OSA) [1].

Some common anatomical factors that can contribute to nasal obstruction or congestion include deviated nasal septum (crooked cartilage within the nose blocks the airway), enlarged turbinate (bony structures within the nasal cavity), nasal polyps (benign growths within the nasal cavity), enlarged adenoids (block the back of the nasal passage) and nasal tumours (benign or cancerous) [1,2]. Nasal obstruction in an infant is usually noted very soon after birth. Nasal obstruction in neonates, who are obligate nasal breathers, can be identified by signs like difficult and noisy breathing, especially during feeding, and cyclical cyanosis that improves with crying and worsens with feeding. Other symptoms include stertor, nasal flaring, apneic episodes, and feeding difficulties [6]. In this case, the baby got lip tie, so she did not get any problem with breathing, because her mouth was not totally closed.

The age of the patient may predict the etiology of nasal obstruction. Congenital defects must be considered when nasal obstruction occurs during the neonate period [1–3,7]. The baby was brought to our hospital with complaints of shortness of breath and wheezing. Initially, the baby was suspected of having pneumonia, accompanied by a suspicion of laryngomalacia. However, it turns out that during its development, there are signs that the baby has a nasal obstruction, namely when it is difficult to insert a suction catheter to suck out secretions from the nose, where even the smallest catheter tube cannot enter. Even though the baby was 10 days old and there was no information from the health workers who handled her before the baby was suspected of having a congenital abnormality in his nose.

On rhinoscopy, it was found that there was a blockage of the choana, so the ENT doctor did not dare to insert the catheter further for fear of rupture. So, a nasal endoscopy was performed, and it was found that there was an elevation of the palate durum covering the choana. Given this fact, it was necessary to explore further the cause of this elevation of the palate durum. Based on the facial CT performed later, it was found that the blockade of the nasal choana originated from the meningocele, originating from a defect in the basal skull. So, it finally became clear that the cause of nasal obstruction in this case was the basal meningocele covering the nasal choana. Airway obstruction in a neonate may be caused by the abnormal development of typical airway structures (e.g. choanal atresia and pyriform aperture stenosis) or by a mass obstructing the airway. There are several causes for a nasal mass in a neonate. These include nasal dermoid cysts, gliomas, cephaloceles, and rarer entities such as benign

fibrous histiocytomas and hamartomas. [1–3,5] Due to the typical obligate nasal breathing pattern seen in neonates, any cause of nasal obstruction may present with features of respiratory distress. When a nasal cavity mass contains neural tissue, the three important differential diagnoses are cephalocele, nasal glioma and teratoma. A cephalocele is an abnormal protrusion of the brain or its coverings. It may contain brain tissue (encephalocele) or solely meninges (meningocele). Nasal gliomas are like cephaloceles but have lost their intracranial connection. Teratomas may be differentiated by examining the entire specimen for germ cell layers [1]. Encephaloceles have been reported as a cause of nasal mass in newborns and are usually described with typical associated features such as cleft lip and hypertelorism [4,8,9].

Nasal encephaloceles are crucial because they can present with CSF rhinorrhea or nasal obstruction in children. To date, the etiology is unknown but hypothesis includes localized weakness in the bony structure of the face associated with factors such as hyperthermia, viral infections, or folic acid deficiency during pregnancy [4]. In this case, the etiology of basal meningocele was unknown because there was no sufficient data.

This case is interesting because the diagnosis was made through a long journey, where the nasal obstruction was not detected from the beginning of birth. This is a warning to all health workers who help deliver babies so that after the baby is in stable condition, a thorough examination is carried out so that it can detect congenital abnormalities early. It is a standard procedure that every newborn should have their nostrils checked to ensure there is no problem [9]. A nasal catheter insertion should be done to explore the cause if nasal obstruction is suspected. Upper respiratory tract obstruction etiology in early life can be a life-threatening anomaly, as in this case.

A case series reported before was described to emphasize the great importance of investigating the state of the nasal airways in any newborn infant who has apparent difficulty breathing [3]. It is simple to pass a soft rubber catheter, lubricated with a little paraffin, back through each nostril; this may relieve cyanotic symptoms and indicate a diagnosis when obstruction is present. If the nasal obstruction cannot be overcome in this simple way, then it is essential that the mouth be kept open and an airway is maintained through this route until such time, generally about ten days, as the baby learns to breathe through the mouth. Feeding will be difficult during this period since suckling is impossible, and it must therefore be maintained using a spoon or in very ill babies by the passage of an orogastric tube two or three times a day.

In this case, the baby came to the hospital with the main complaint of shortness of breath and stridor. According to the results of the anamnesis and physical examination, as well as X-ray photos, the patient was diagnosed with pneumonia. This fact had covered up the leading cause of her breathing difficulty and postponed its origin etiology investigation. Aspiration pneumonia in lip tie and atresia choana case is a usual complication due to the baby's poor coordination of breathing and drinking system [1,6].

Meningitis that occurred early in life in this patient is thought to be due to external exposure to the cerebrospinal fluid, resulting in bacterial infection. External exposure, in this case, is due to cerebrospinal fluid leakage from the base skull defect.

There are two methods of handling meningocele in this case: the nasal or cranial approaches [6,10–12]. Pediatric meningoencephalocele has been traditionally addressed with transcranial approaches which have obviously higher complication rates. Transnasal endoscopic repair of anterior skull base meningoencephalocele is a minimally invasive single stage surgery, and has advantage in terms of lesser hospital stay, cost of treatment, and better cosmesis [6,12]. The repair technique should be tailored to the size of defect to provide a water-tight seal for better outcome [6]. However, in Surabaya, there are still no facilities to perform surgery through the nasal approach, so in this case, surgery was performed through a cranial approach performed by a neurosurgeon.

Untreated children may present with recurrent meningitis and/or epilepsy not controlled by medications [4,5]. The baby got meningitis 5 months later after getting a severe cold. This is due to the leakage of the surgical wound in the defect area, resulting in meninges

rupture. The cerebrospinal fluid leakage through the surgical wound defect is indeed at high risk if there is an increase in intranasal pressure due to an intense cold [3,4]. Although the leakage could be repaired, the complication of meningitis in the form of blockade of the ventricular cavity was unavoidable, resulting in hydrocephalus and the baby was fitted with a ventricular-peritoneal shunting. It is necessary to avoid severe colds to prevent recurrent surgical wound leakage [5,6].

CONCLUSION

Every newborn must undergo a thorough physical examination to detect airway obstruction especially nasal obstruction. In our case, the nasal obstruction was not detected from the beginning of birth, and it led to delays in finding the cause. Basal meningocele in this case was diagnosed accidentally based on a facial CT scan that explored the causes of choana atresia. It's essential to detect choana atresia since birth, explore the etiology immediately, and manage it well to prevent life-threatening complications.

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