CASE REPORT

Nasopharyngeal encephalocele: a rare cause of upper airway obstruction

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Nasopharyngeal encephalocele is a rare, benign congenital anomaly. It has the potential to be fatal due to airway obstruction. Here, we report on a 34-day-old infant with pneumonia who underwent mechanical ventilation. An upper airway evaluation was performed due to prolonged intubation, and revealed the presence of a nasopharyngeal encephalocele. The patient tolerated extubation and oral feeding after surgical resection of the lesion. Awareness of the condition can help clinicians arrive at an earlier diagnosis and enhance management.

Introduction

Nasopharyngeal encephalocele is a rare, benign congenital anomaly. It has the potential to cause fatal airway obstruction. Awareness of the condition can help clinicians arrive at the diagnosis earlier and enhance management. Here, we report on a 34-day-old patient with a nasopharyngeal encephalocele who developed respiratory insufficiency treated by mechanical ventilation secondary to airway obstruction.

Case report

This 34-day-old ex-35 weeks preterm female infant was born to a mother with triplets. She was admitted to our intensive care unit from another hospital for respiratory distress due to pneumonia in May 2008. The pregnancy had been normal and delivery was uncomplicated. Prior to this admission she was diagnosed to have neonatal pneumonia deemed to require mechanical ventilation. The patient had had a suction catheter passed through her nostrils with some difficulty as part of regular screening for the presence of choanal atresia. The patient received a course of antibiotic therapy in our intensive care unit. Despite improvement in the chest X-ray, her clinical condition did not stabilise and therefore we repeated upper airway examination. After unsuccessful attempts to pass the suction catheter through the nostrils, she underwent paranasal computed tomography (CT). This demonstrated a hypodense mass lesion with solid and cystic components extending from the left nasal cavity through the nasopharynx. Magnetic resonance imaging (MRI) further delineated the lesion's dimensions as 13 x 17 mm with well-defined borders and without any communication with cerebrospinal fluid (Fig 1). After an endoscopic examination, the

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Airway obstruction; Encephalocele; Respiratory insufficiency

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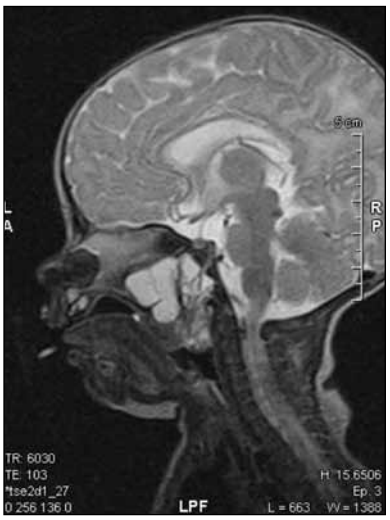

FIG 1. A T2-weighted sagittal magnetic resonance image demonstrating nasopharyngeal hyperintense mass lesion without intracranial connection

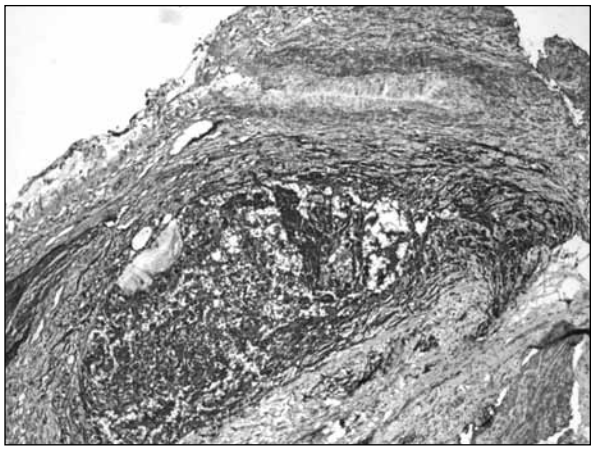

FIG 2. Positive immunostain for glial fibrillary acid protein showing areas of glial tissue beneath the respiratory epithelium (DAB chromogen, x 100)

mass lesion was excised under general anaesthesia. The patient was extubated on postoperative day 1, and pathology confirmed the diagnosis of an encephalocele (Fig 2).

Discussion

Nasopharyngeal masses may lead to neonatal airway obstruction such as those occur in choanal atresia, since infants are obligate nasal breathers for the first few months of life.[1] All newborns should routinely be screened for nasal obstruction by transnasal passage of suction catheters to the oropharynx.

In our patient, presence of initial passage of the catheter, though with some difficulty, delayed the diagnosis and repeat examination was performed only after persistent respiratory distress despite the recovery from pneumonia. Not only failure but also difficulty in passing a nasal catheter should have alert clinicians to the possibility of nasal obstruction.

A nasal encephalocele is a rare lesion, which may present with upper airway obstruction in the neonatal period.[2] Proposed mechanisms for its pathogenesis include failure of the ossification of the skull base, or a failure of the anterior neuropore to close by the end of the fourth week of gestation.[3,4] Imaging should include both CT and MRI.[5] The CT is superior for defining bony deformities and MRI is better for revealing any intracranial connection. Early surgical resection is the mainstay of management, as relieving the obstruction allows extubation and oral feeding.[6] Whilst this condition is rare, we would like to re-emphasise the importance of repeat upper airway examinations in patients with unexplained respiratory distress, particularly as this condition may well be overlooked when the clinical picture is associated with pneumonia.

鼻咽腦膨出：一種罕見的上呼吸道阻塞的原因

鼻咽腦膨出是一種罕見的良性先天性異常病。此病可能會引致氣道阻塞而致命。本文報告一名34天大患有肺炎的嬰兒接受機械性通氣。嬰兒需長期插管，為她檢查上呼吸道時發現有鼻咽腦膨出的情況。手術切除病變後替病人拔管，及後她可以進食。了解此病可幫助醫生盡早作出診斷及處理病情。

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