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Case Report

Child with intranasal meningoencephalocele within the middle turbinate: case report

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ABSTRACT

Cephalocele or encephalocele consists of the herniation of cranial contents through a bony defect in the skull. An occipital meningoencephalocele is the most common whereas an intranasal meningoencephalocele is a rare type of cephalocele. Here, we report a case of an intranasal meningoencephalocele within the middle turbinate. A 9-year-old male child was admitted to the emergency department due to epistaxis and difficulty breathing. Computed tomography (CT) and magentic resonance imaging (MRI) results revealed herniation of a voluminous, predominantly cystic lesion next to the cribriform plate, extending into the right middle conus. During endoscopic endonasal surgery, a bulging middle turbinate was visualized and the herniated sac was found to be inside the turbinate. The herniated sac, together with the middle turbinate, was resected. The patient presented good evolution, with no signs of cerebrospinal fluid fistula. Intranasal meningoencephalocele is rare. Usually, the herniation is between the middle turbinate and the nasal septum, unlike in this case, where it was within the middle turbinate itself. Clinical symptoms can vary and include nasal obstruction, anosmia, rhinorrhea, epistaxis and meningitis. Imaging studies (CT and MRI) help with differential diagnosis (polyps, nasal glioma, dermoid cyst, teratoma, mucocele) and surgical planning. Surgical treatment can be open or endoscopic, as in the case reported here. In children with nasal masses, especially when there are no signs of cerebrospinal fluid fistula, intranasal meningoencephalocele should be considered, thus avoiding puncture or biopsy.

Keywords: Encephalocele, Intranasal, Endoscopy, Meningitis

INTRODUCTION

A cephalocele, or encephalocele, consists of the herniation of cranial contents through a bony defect in the skull. This herniated content may consist of meninges only, called a meningocele, or it may be meninges with brain tissue, called a meningoencephalocele.[1] The classification is based on the location and type of skull defect. Encephalocele of the skullcap is the most common, occurring in 80% of cases in the occipital area. Therefore, intranasal meningoencephalocele is rare and herniation of the neural elements through the cribriform plate usually occurs between the middle turbinate and the nasal septum.[2,3] The report presented here is of a rare case of intranasal meningoencephalocele within the middle turbinate.

CASE REPORT

A 9-year-old male child was admitted to the emergency department due to epistaxis and difficulty in breathing; symptoms that worsened 30 days prior to admission. A 9-

year-old male child complained of nasal obstruction since birth, with sporadic bleeding. Child did not have any trauma, allergy symptoms, or previous surgeries.

Rhinoscopy showed a soft mass in the right nasal fossa with almost complete obstruction and septal deviation to the left, with no abnormalities in the nasopharynx. Computed tomography (CT) of the paranasal sinuses showed a large, predominantly cystic expansive lesion implanted in the roof of the nasal cavity, causing deviation of the septum and with no apparent bone erosion. Magnetic resonance imaging (MRI) showed herniation of part of the brain tissue (meninges) next to the cribriform plate in the anterior portion, extending into the middle turbinate (Figure 1).

Endonasal endoscopy revealed a bulging middle turbinate (Figure 2A) and it was found that the herniated sac was inside the middle turbinate. This was followed by cauterization and resection of the herniated sac together with the middle turbinate (Figure 2B and D). Subsequently, fat and a hemostatic sponge were inserted into the defect in the cribriform plate (Figure 2C). The patient presented good evolution, with no signs of cerebrospinal fluid fistula, and he was discharged from hospital on the fifth postoperative day.

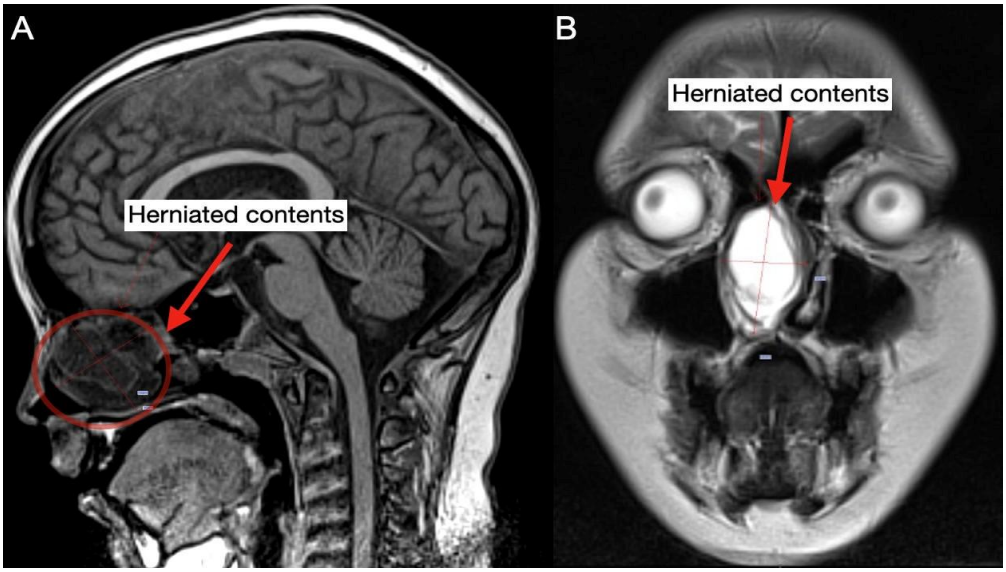

Figure 1: (A) Sagittal t1-weighted MRI showing the herniated contents of the roof of the nasal cavity; (B) coronal t2-weighted MRI showing the herniated contents within the middle turbinate.

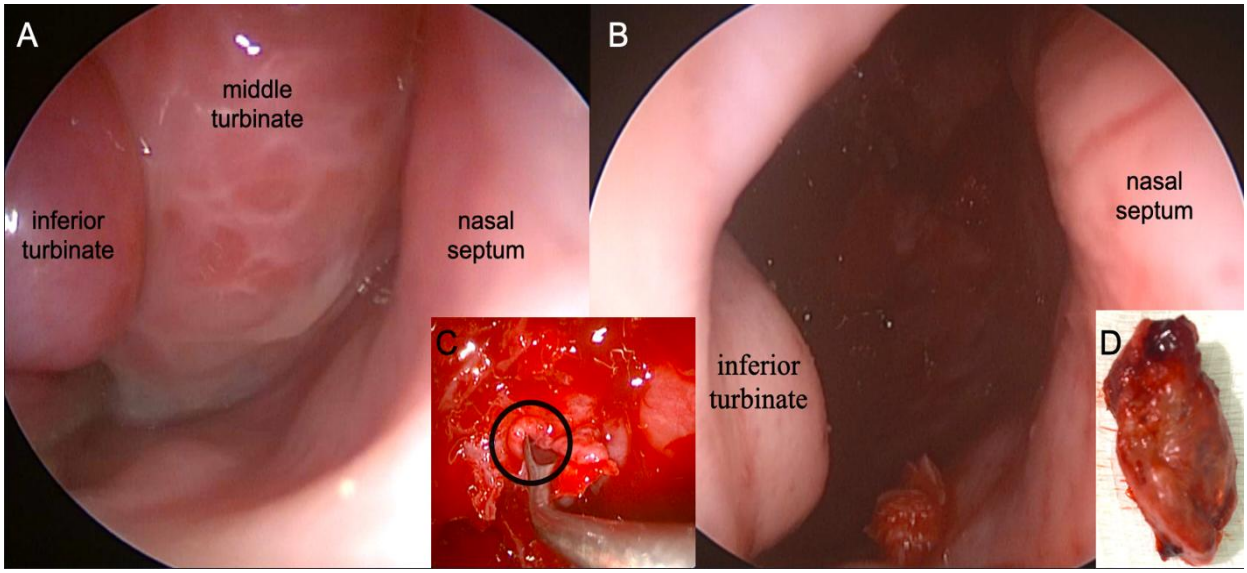

Figure 2: (A) Herniated sac in the middle turbinate; (B) the middle turbinate was resected along with the meningoencephalocele; (C) region of the cribriform plate defect where herniation occurred; (D) middle turbinate removed.

DISCUSSION

Intranasal meningoencephalocele is not very common. The etiology may be congenital, traumatic, or spontaneous. In congenital cases, it may be due to neural tube disorders in the 3rd or 4th week of development or disorders in the development of the cranial bones. The congenital defect is more common in children, whereas traumatic or spontaneous origin is predominant in adults.[4,5] Jabre et al consider that congenital encephalocele in adults is rare because facial abnormalities are subtle or absent.[6] When there is no defined cause, the encephalocele is called spontaneous. McPheeters et al state that several apparently spontaneous encephaloceles may actually be post-traumatic, due to events that the patient does not remember or considers irrelevant.[7]

The patient may experience a variety of symptoms, such as nasal obstruction, anosmia, rhinorrhea, epistaxis, and even meningitis. The diagnosis is more obvious when the child presents with constant rhinorrhea with a unilateral nasal mass. Complaints of headache and a history of head trauma are also relevant.[4-8] If a nasal mass present, differential diagnoses are important, including those of nasal glioma, dermoid cyst, teratoma, nasal polyps, and nasal mucosa cyst.[3] In the case reported here, the clinical picture was of nasal obstruction and epistaxis, with no history of trauma. Rhinoscopy revealed a unilateral nasal mass with septal deviation.

CT is generally used as a diagnostic method to study the bony anatomy of the skull base, complemented by MRI showing details of the herniation of brain tissue into the nasal fossa.[3] In the report described, CT showed the implantation of the lesion in the nasal cavity, its relationship with the bony structures, and the degree of erosion-type involvement. MRI showed details of the contents of the herniated sac (part of the brain tissue) inside the middle turbinate.

The treatment for intranasal meningoencephalocele is surgical. The case should be studied carefully and punctures or biopsies should be avoided, as these cannot solve the problem and there is a possibility of infections such as meningitis.[3] Therefore, the approach can be taken via the conventional open route with a frontal craniotomy, separating the cranial portion from the nasal portion, and the bone defect can be reinforced with fascia lata, or muscles, or bone fragments. In contrast, endoscopic approach, when performed by experienced surgeon, reduces trans-operative time and surgical morbidity.[8] The best treatment option in the case described was the endoscopic approach, in which the herniated sac was visualized inside the middle turbinate followed by definitive resolution of the case.

CONCLUSION

Intranasal meningoencephalocele is rare, especially when the herniated neural content extends into the middle turbinate. It is important to remember that nasal masses may be a case of meningoencephalocele in children, even if the masses do not show signs of cerebrospinal fluid fistula. It is not advisable to perform punctures or biopsies; rather, opting for definitive treatment such as endoscopy is advised.

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