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CASE REPORT

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Congenital intranasal meningocele in a newborn – case raport

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Summary

Background: Meningocele or meningocephalocele is a rare defect, which consists in the displacement of the structures of the brain or/and meninges through a congenital cavity in the cranium. Depending on the location, there are hernias of the skull vault and base or external and internal hernias. Some of them can cause breathing disorders in children.

Case Report: We described a case of an eutrophic, full-term newborn boy with breathing disorders. A laryngological examination found a tumor of the left nasal meatus, from which a sample was collected for histopathological study. Due to a cyst revealed under the lesion and pulsation of the tumor, it was decided to include imaging examinations into the diagnostic process. Computed tomography (CT) showed a mass of meningocele type, which was confirmed in magnetic resonance imaging (MRI).

Results: In case of breathing disorders and a suspected mass in the nasal meatus in a newborn, a congenital meningocele or menigocephalocele should be assumed. This constitutes a contraindication to sampling of the lesion, without prior imaging tests (CT and/or MRI).

Key words: MRI • CT • newborn • congenital intranasal meningocele

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Background

Menigocephalocele is a dislocation of the brain structures and/or meninges through congenital bone defects of the skull. Depending on the content of the hernial sac, the hernias can be classified into meningoceles and menigocephaloceles, and depending on the location of the ring of hernia within the skull – into hernias of the base and vault. Hernias may be external and internal ones. Most of the hernias of the vault are found in the occipital area. Among hernias of the skull base there are:

1. frontal-ethmoidal ones, including a) naso-frontal, b) naso-ethmoidal, and c) naso-orbital,
2. sphenoid-orbital,
3. aphenoid-maxillary,
4. naso-pharyngeal.

The aetiology of the meningoceles is not clear. There are two most popular theories. One of them mentions the disorders of embryogenesis of the neural tube in the 3rd–4th week of development. The second one focuses on the disorders of development of the cranial bones [1–3].

The highest diagnostic problems are caused by hernias of the skull base, especially the internal ones, as they do not deform the face [2,4]. They are often misdiagnosed as polyps or tumors [5]. Inappropriate treatment of undiagnosed hernias may lead to a secondary meningitis or fluid leak due their incision or puncture. Hernias protruding to the nasal cavity or nasopharynx may give such symptoms as respiratory disorders, swallowing disorders, cerebrospinal fluid leak or meningitis. Anterior meningoceles and menigocephaloceles belong to rare congenital malformations. Their endemic foci are found in Asia (in Thailand

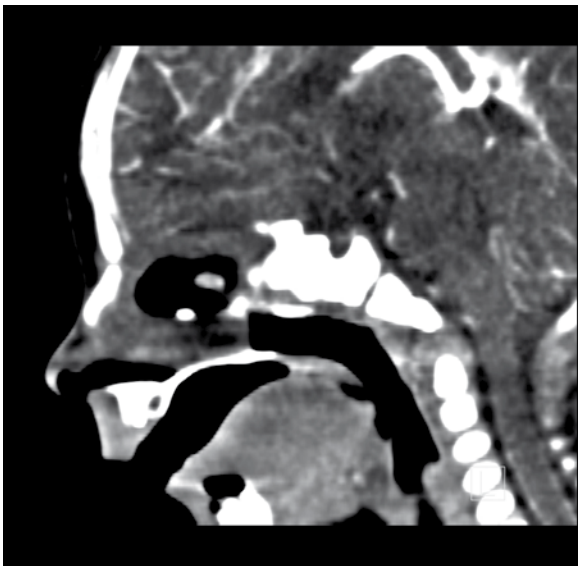

Figure 1. A sagittal contrast-enhanced CT demonstrating a soft tissue mass in the left nasal meatus.

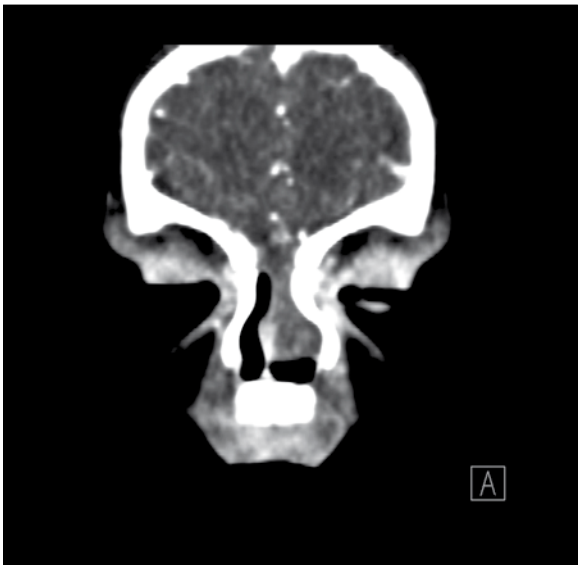

Figure 2. A contrast-enhanced CT – coronal plane. The same case.

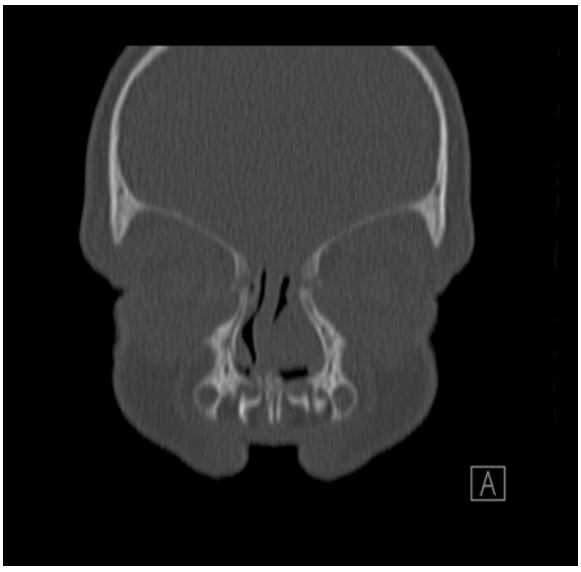

Figure 3. A contrast-enhanced CT – coronal plane. The same case.

Figure 4. Volume-rendering (VR) technique. The same case.

mainly) – incidence of 1:5000 live births. In America and Europe, the incidence of hernias amounts to 1:35000–40000 live births [6–9]. A particularly useful imaging examination in the diagnostics of such cases is the magnetic resonance imaging (MRI) [10]. The aim of this work was to encourage the neonatologists and laryngologists to a thorough analysis of symptoms that may suggest the presence of a meningocele or menigocephalocele in the nasal cavity, imitating polyps or tumors. This should lead to the introduction of a proper diagnostic or therapeutic process.

Case Report

An eutrophic, two-week, full-term male newborn was admitted to the Department of Otolaryngology of the Clinical Hospital in Poznań with a suspected tumor of the left nasal meatus. In the first days of his life, his mother observed breathing difficulties. On physical examination, the physician found a soft-tissue mass in the left nasal meatus, and made an initial diagnosis of angioma.

Anterior rhinoscopy found a lesion mass protruding from the vault of the nasal cavity, on the left side of the septum. A sample was collected for histopathological examination. A cystic lesion with pulsation was found at the base of the solid part of the tumor. A clear fluid was aspirated. No pathological lesions were found on endoscopy of the oral cavity, pharynx and ears. The results of the laboratory tests were normal. A CT of the facial cranium showed a soft-tissue area in the left nasal meatus, with inhomogeneous absorption rate, ranging from -9 to 50 HU. The area was of 3.1×0.9×0.9 cm in size (ap×ds×cc) and did not reveal a significant enhancement after contrast medium administration. The lesion was displacing the nasal septum to the right and constricting the right nasal opening. No features of a tumor or symptoms of osteolysis were found. Lymphatic angioma could not be excluded (Figures 1–4).

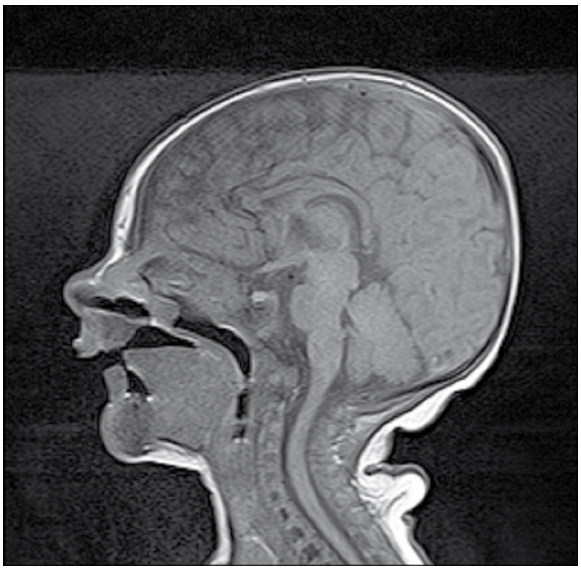

Figure 5. A sagittal T1-weighted image. The same case.

Figure 6. A sagittal T1-weighted image. The same case.

The continuity of the lesion with structures of the brain in the anterior cranial fossa was the cause of further diagnostics, including the MRI in SE, TSE, and FLAIR sequences, in axial, sagittal, and frontal planes, and in T1- and T2-weighted images. A menigocephalocele was found in the left nasal cavity, with hypoplasia of the nasal meatuses in the left nasal duct, as well as very wide semicircular ducts, bilaterally. The MRI image of the brain was normal otherwise (Figures 5–7).

The patient was qualified for surgery which was carried out by a neurosurgeon and otolaryngologist. Plastic surgery of the left naso-ethmoidal menigocephalocele was performed with craniotomy, from the left frontal approach, through the midline. Crista galli was exposed and partially excised in order to show the site of protrusion of the basal hernia through the left ethmoid bone. After an incision of the dura mater, the herniated brain tissue was cut off, and the meninx around the defect was dissected. At this stage,

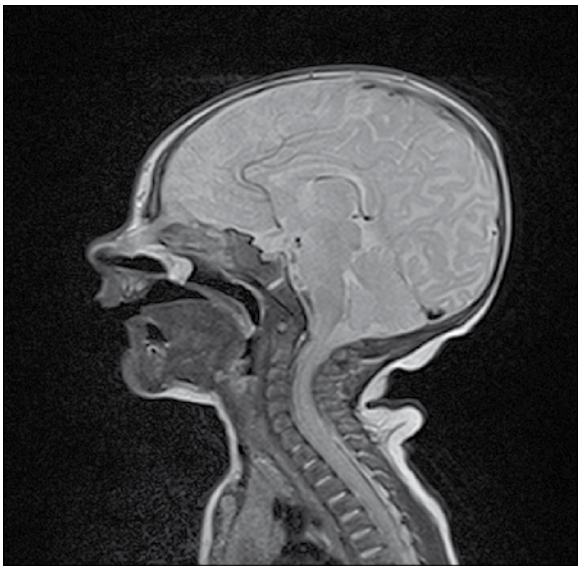

Figure 7. A sagittal T2-weighted image. The same case.

the laryngologist removed the intranasal part of the hernia. The removed brain tissue was subjected to histopathological examination. Nasal cavity inspection by laryngologist confirmed that the whole pathological tissue was removed. The defect in the ethmoid bone was sealed with tachosil patches. Dura mater was sewn. Bone flap was put back in place and the skin was sewn. After an uncomplicated postoperative period, the child was discharged home in a good general health state. The patient remains under the care of a physician from Neurosurgery Clinic and one from Otolaryngology Clinic. Further examinations, carried out after one month following the procedure and including laryngological and neurosurgical tests, found no deviations from normal values.

Discussion

Menigocephaloceles may be located at many sites, leading to different symptoms i.e.: deformations of the facial skeleton, visual disturbances, disturbed nasal patency, disturbances of smell, cough not accompanied by any symptoms of infections of the respiratory tract, or meningitis. Most of these symptoms are reported by adults, during history taking. Additional diagnostic difficulties, especially in pediatric cases, are connected with the fact that the observed symptoms start to affect people around, and especially parents. A basic symptom of hernia in children, located in the nasal cavity, is breathing difficulties, which were also found in our case. Anterior rhinoscopy showed a mass lesion protruding from the vault of the nasal cavity. A sample was collected for histopathological examination. Due to the observed pulsation, further surgical procedure was abandoned. According to the literature, an absolute contraindication to diagnostic sampling is even the slightest risk of menigocephalocele. Differential diagnosis of this disorder should include: nasal polyps, cysts, mucocele, tumors, and allergic rhinitis. Diagnostic examinations use the MRI and CT method. Non-calcified parts of the skull cause many difficulties with CT interpretation. In such cases, MRI is particularly useful. It provides images with high contrast among different soft tissues and with high resolution in

any plane, and it allows for elimination of artifacts from bone structures [11]. Therefore, in the described cases, MRI is a basic examination method differentiating mucoceles and menigocephaloceles from nodular lesions of a different type. However, there still exists the problem of its availability at many diagnostic centers. What is more, the specificity of patients' age requires a cooperation of an anesthesiologist. Summing up, widely applied diagnostic tests with CT should take into account this rare location of congenital menigocephaloceles, i.e. nasal cavity. Due to such a location, the menigocephaloceles require also an extensive cooperation of neurosurgeon and otolaryngologist, so that an optimal surgical method could be chosen [12].

Conclusions

1. In children with breathing difficulties, who were diagnosed with tissue masses in the nasal cavity, menigocephaloceles or meningoceles should also be suspected.
2. Menigocephaloceles on the floor of the anterior fossa, located in the nasal cavity, are an absolute indication to sampling for histopathological examination.
3. MRI is a noninvasive, safe imaging method which allows (as opposed to CT) for a definite diagnosis of menigocephaloceles and meningoceles.
4. Treatment of meningoceles and menigocephaloceles requires surgeries attended by a neurosurgeon and otolaryngologist.

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