A case of idiopathic encephalomeningocele

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Abstract

In the present case we report about an encephalomeningocele in an adult female. Since the cause of this medical entity is a congenital fusion defect of the neural tube of the cranial base, most of the encephaloceles occurs in children leading to facial disfigurement. In the rare cases described in adults, rhinorrhea is usually present. Here we present a case of temporobasal encephalomeningocele in a 72-year-old female patient suffering from headaches in the last 4-5 years. No rhinorrhea or other significant neurological symptoms were noticed. No congenital cause was apparent. After diagnostic steps including brain magnetic resonance imaging (MRI), cranial computed tomography (CT) and MR cisternography, an encephalomeningocele was diagnosed. Through a pterional approach this was completely removed. The only symptom the patient complaint about, headache, was eliminated after surgery.

Introduction

Encephalomeningocele is a herniation of the brain and meninges through a congenital bone defect resulting from the failure of normal midline fusion of the cranial neural tube.[1] It is an uncommon clinical entity occurring in less than 1/35,000 persons.[2] In Caucasians, more than 70% of the encephalomeningoceles are located on the occipital/posterior part of the brain, however various sites are possible. As congenital disorders, they are usually diagnosed in childhood as a result of midfacial anomaly, optic disc anomaly, brain anomaly, cerebrospinal fluid rhinorrhea, recurrent meningitis, visual and endocrinologic disturbance or mental retardation.[3-5] There are only very few cases of encephalomeningoceles in adults.[6,7]

Case Report

In this case we present a 72-year-old female patient complaining of mild cephalgia for the last 4 years localized in the right hemicranium. Further a hypesthesia of the right half of her face was reported. There was a mild disturbance of concentration. No other neurological symptoms could be evaluated. The pain was less severe when treated with analgesics. A magnetic resonance imaging (MRI) has been performed which showed a suspicious mass on the right skull base. She was introduced to the department of Ear, Nose, Throat (ENT) and since the mass did not look like a typical mucocele, admission to our department followed. In the medical history there was no meningitis, encephalitis or congenital anomalies reported. She reported a head trauma in the age of 16-year-old which was treated ambulatory and was not severe.

In the MRI of the brain a multiple septated mass in the right sphenoidal corpus could be identified with migration through the infratemporal area and to the ala major of the sphenoidal bone. The bone of the pterygoid was thinned. In some sections the impression was made that there was a communication of the mass with the subarachnoid space. There was no significant contrast enhancement. An MRI cisternography showed clearly a contact between the mass and the ventricular system. An encephalomeningocele was suspected and an operative treatment with removal of the mass was indicated and offered to the patient (Figure 1).

Surgery was performed through a right pterional approach. Immediately after craniectomy a bluish cystic mass was seen under the excessively thinned bone of the sphenoidal wing. In the temporal area dural degeneration with a penetration was identified and arachnoidea was herniated through the lesioned dura which lead to the encephalomeningocele. The bone of the temporal skull base was very thin and the area where the mass protruded was found. Liberation of adhered brain tissue from the cele followed and the cele was completely removed. Medially of the encephalomeningocele the maxillary nerve was identified. It had contact to it, explaining the hypesthesia the patient suffered from, but was not adherent to it. Inspection of the mass identified cerebrospinal fluid (CSF), some regions were hemorrhagic and others had solid material. The dura was closed with poly-p-dioxanon PDS foil and tissue glue. The space the cele occupied, was filled with abdominal fat (Figure 1). The postoperative MRI of the brain showed a complete removal of the histologically proved encephalomeningocele. The patient was discharged 10 days later. Headaches and trigeminal hypesthesia were considerably better.

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Discussion

In this case we present a 72-year-old female patient with a big temporobasal encephalomeningocele that was diagnosed in a routine imaging control because of headaches. Her neurological status was almost intact. Headaches were the main symptom. In the majority of celes described in adults rhinorhea was present.[3]

The preoperative diagnostic procedures included MRI brain scans, thin-sliced CT of the cranial base and MRI cisternography and are similar to that described in other studies. A clear communication with the CSF space was noticed; therefore the existence of an encephalomeningocele was suspected. None of the known causes like congenital or infectious were present, therefore the present case is one of an idiopathic and rare encephalomeningocele.

The treatment of choice was operative. Through a pterional osteoplastic craniotomy the cystic mass was completely removed and the skull base reconstructed. Surgery is the only alternative for definitive treatment of encephalomeningoceles.[1] Depending on the location, different procedures have been performed. Most of the celes occur in children[7] and rarely there are cases describing encephalomeningoceles in adults.[6] The majority of encephalomeningoceles are located in the frontobasis and treatment needs a transfacial approach resulting in facial disfigurement. The frontoethmoidal type as an example can be cured through a bifrontal

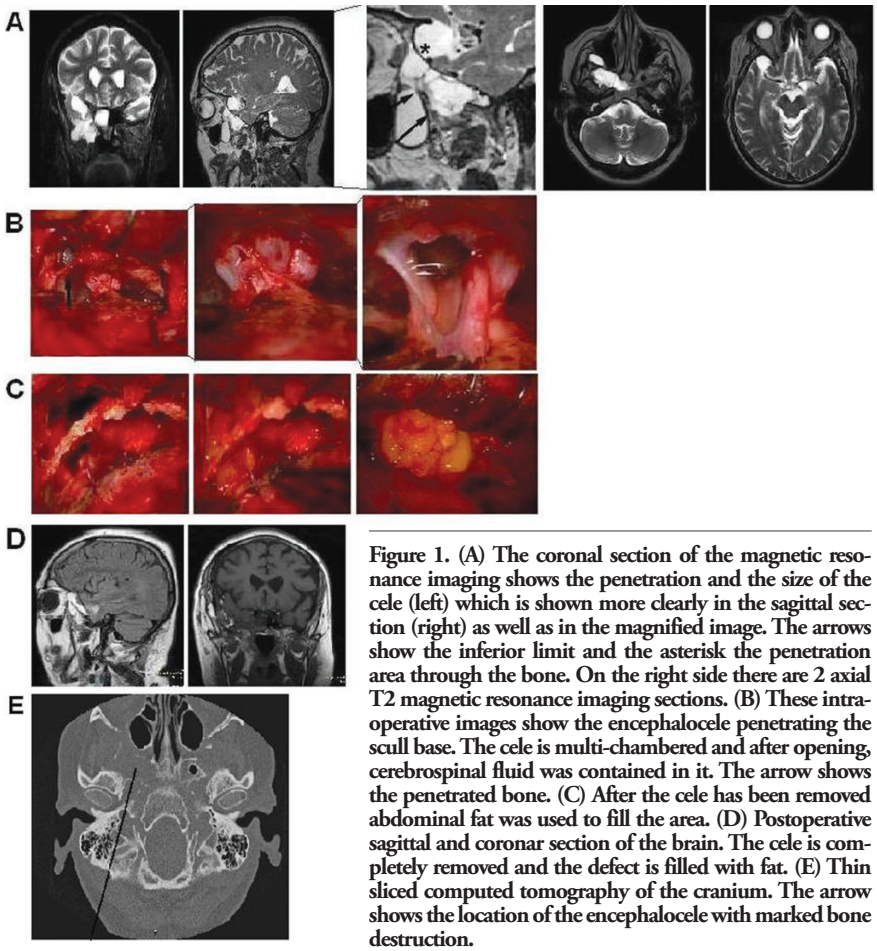

Figure 1. (A) The coronal section of the magnetic resonance imaging shows the penetration and the size of the cele (left) which is shown more clearly in the sagittal section (right) as well as in the magnified image. The arrows show the inferior limit and the asterisk the penetration area through the bone. On the right side there are 2 axial T2 magnetic resonance imaging sections. (B) These intraoperative images show the encephalocele penetrating the scull base. The cele is multi-chambered and after opening, cerebrospinal fluid was contained in it. The arrow shows the penetrated bone. (C) After the cele has been removed abdominal fat was used to fill the area. (D) Postoperative sagittal and coronar section of the brain. The cele is completely removed and the defect is filled with fat. (E) Thin sliced computed tomography of the cranium. The arrow shows the location of the encephalocele with marked bone destruction.

craniotomy and dural plastic for the CSF rhinorrhea.[6] In the present case a common pterional approach was performed so that no facial disfigurement occurred. The location of the cele allowed such an approach.

Conclusions

Here we present another rare case of encephalomeningocele in an adult which occurred without apparent etiologic cause in an atypical area in the skull and the appropriate treatment method.

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