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Case Report

Arachnoid cyst with extraordinary extracranial extension in the skull base as a result of an iatrogenic defect of the middle cranial fossa floor: Case report and literature review

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SUMMARY: **Arachnoid cysts are benign intracranial lesions that are typically diagnosed incidentally. They are divided into two types: congenital and acquired. Acquired arachnoid cysts are rare and usually arise after trauma, infection, or haemorrhage. In this report, a rare case of an iatrogenic multiloculated arachnoid cyst as an unusual complication of a skull base defect is presented. It extended extracranially into the sphenoid sinus, the ethmoid, the infratemporal fossa, the nasopharynx and the nasal cavity, as well as into the pterygomaxillary and retromaxillary space, appearing with a distinct clinical picture. We discuss the differential diagnosis and the potential causes of the lesion and provide a brief review of the literature.** © **2009 European Association for Cranio-Maxillo-Facial Surgery**

Keywords: **arachnoid cyst, sphenoid sinus, skull base defect, middle cranial fossa, mucocele**

INTRODUCTION

Arachnoid cysts are intra-arachnoid collections of cerebrospinal fluid (CSF) (*Rengachary* and *Watanabe*, 1981; *Catala* and *Poirier*, 1998). They are mostly discovered incidentally in patients with minor head trauma, headache, or symptoms from another lesion. Symptoms can develop as cysts grow and exert mass effects on surrounding structures. Symptoms include craniomegaly, localized cranial bulging, signs of increased intracranial pressure, seizures, focal neurological deficits, psychomotor retardation and endocrine disorders (*Di Rocco*, 1990; *Pierre-Kahn* et al., 1990; *Ciricillo* et al., 1991; *Venes and Brunberg*, 1993; *Rengachary and Kennedy*, 1996). However, many cysts remain asymptomatic, and thus their treatment remains controversial. Although they are regarded as histologically benign, developmental abnormalities of the arachnoid membrane (*Di Rocco*, 1990; *Pierre-Kahn* et al., 1990; *Venes* and *Brunberg*, 1993), arachnoid cysts that develop postoperatively or postraumatically have also been reported (*D'Almedia and King*, 1981; *Brackett* and *Rengachary*, 1982; *Sato* et al., 1983; *Weaver* et al., 1996; *Choi* and *Kim*, 1998). In this report, a rare case of an iatrogenic multiloculated arachnoid cyst of the middle cranial fossa extending extracranially and appearing with a distinct clinical picture, as an unusual complication of a skull base bony defect, is presented. We discuss the differential diagnosis and the potential causes of this rare lesion and provide a brief review of the literature.

CASE REPORT

A 40-year old man was admitted to our department with a 14-month history of progressively worsening headaches and difficulty in breathing. Twenty-two years earlier the patient had undergone a removal of a nasopharyngeal angiofibroma, extending into the middle cranial and the infratemporal fossae. The tumour was removed at that time by an infratemporal approach Fisch Type C and the dural defect was patched with facia lata and with a piece of free muscle flap. Two years later a mucocele of the sphenoid sinus was diagnosed and removed via a lateral rhinotomy approach. The further postoperative course was uncomplicated for 20 years without any sign of recurrence. 14 months before his present admission, the patient started complaining about progressively worsening headaches and difficulty in breathing. Endoscopy of the nose showed obliteration of both nasal cavities, by cystic masses, covered with normal looking mucosa (Fig. 1).

Magnetic resonance imaging (MRI) showed multiple cystic lesions with signal characteristics similar to that of CSF in the right middle cranial fossa protruding through the bony skull base defect into the infratemporal fossa, the sphenoid sinus, the ethmoid, the nasopharynx, the nasal cavities as well as in the pterygomaxillary and retromaxillary space (Fig. 3). Since communication of the cysts with the subarachnoid space was suspected but mucoceles could not be ruled out, we performed a CT-cisternography for further evaluation of the lesions.

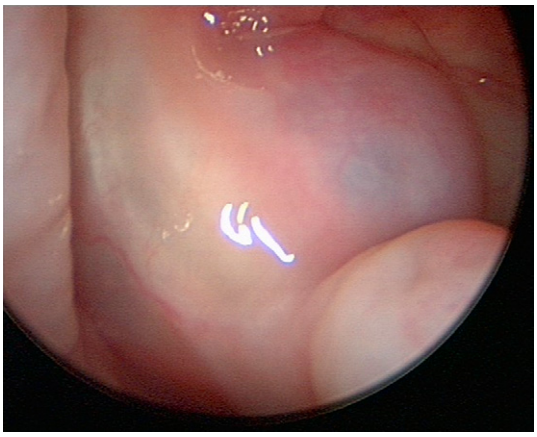

Fig. 1 – Endoscopic image of the left nasal cavity showing a cystic mass, originating from the area of the nasopharynx and the sphenoid sinus. The cyst obliterates the nasopharynx and is covered by normal looking mucosa.

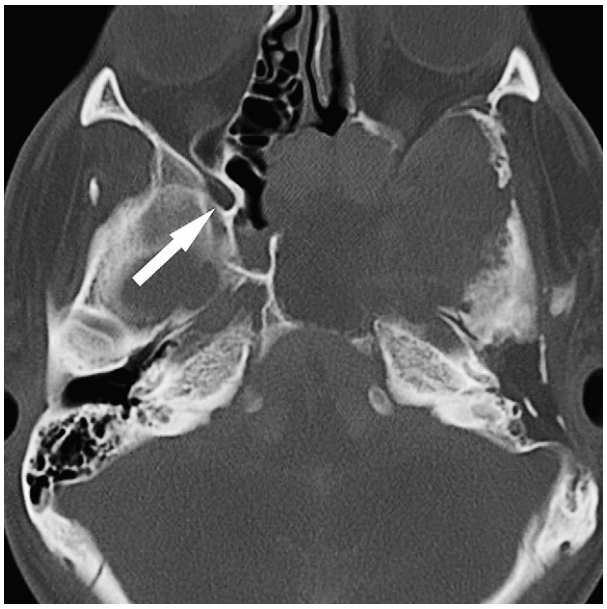

Fig. 2 – Axial CT of the skull base. Note the destruction of nearly the entire left skull base, affecting not only the pterygopalatine fossa (white arrow indicates the normal right), but the apex of the petrous bone, including the carotid canal.

CT-cisternography revealed contrast enhancement in parts of the cystic lesions and arachnoid cysts were diagnosed (Figs. 2 and 4). A left pterional craniotomy was performed. Multiple cystic lesions extending from the floor of the middle fossa to the infratemporal fossa were identified protruding through the dural and bony defect of the sphenoid bone into the sphenoid sinus and the nasopharynx. These were excised. The cysts contained fluid resembling CSF. A defect of the dura mater was observed while the arachnoid membrane was intact. A temporalis muscle flap was used to cover the bony skull base defect. Subsequently an endonasal approach was performed for complete removal of the cysts from the sinonasal cavities. Histological examination of the specimen revealed a cyst wall resembling normal arachnoid. The

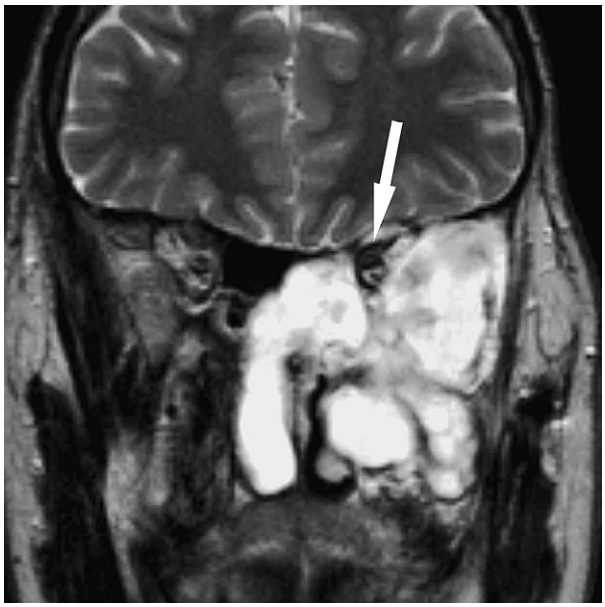

Fig. 3 – T2-weighted (T2W) coronal view at the level of the orbital apex, demonstrating the CSF-containing cysts (bright signal) not only in the ethmoid and nasal cavity, but medial to the left orbit (white arrow) and (inferiorly) in the pterygomaxillary and retromaxillary space.

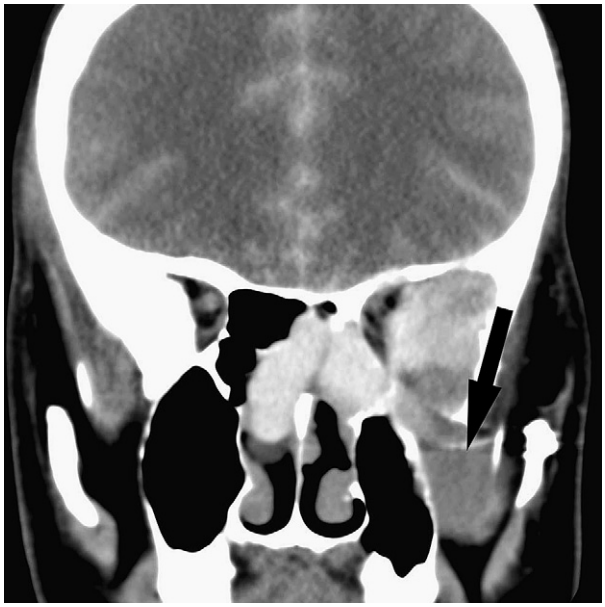

Fig. 4 – Corresponding coronal CT-cisternography, where the arachnoid cysts in the ethmoid, nasal cavity and middle fossa are seen in direct communication with the CSF-space, while the cyst in the pterygomaxillary space (black arrow) shows no direct communication.

postoperative course was uneventful. At the 12-month follow up examination, the patient reported that the difficulties in breathing and headache disappeared. Control imaging studies showed no recurrence.

DISCUSSION

Arachnoid cysts are intra-arachnoid collections of CSF covered by arachnoidal cells and collagen. Primary