

Spontaneous Encephalomeningocele: A Case Report



Heloisa dos Santos Sobreira Nunes*, Eduardo Amaro Bogaz, José Antônio Pinto, Kelly Abdo and Andréia Vasconcelos

Department of Otolaryngology, Núcleo de Otorrinolaringologia de São Paulo, Brazil

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*Corresponding author: Heloisa dos Santos Sobreira Nunes, Department of Otolaryngology, Núcleo de Otorrinolaringologia de São Paulo, Brazil, Tel: +55 11 55731970; Email: helo2005@hotmail.com

Abstract

Herniation of meningeal and brain tissue into the middle ear and mastoid through a bony defect that allows meningeal and/or brain tissue to protrude into temporal bone is a rare pathological entity with potentially life-threatening complications, requiring surgery [1,2]. While herniation of brain tissue in middle ear usually occurs from the middle fossa through a defect either in tegmen tympani or in the mastoid tegmen [1,2], the etiology of this defect varies and includes chronic otitis media with or without cholesteatoma, and previous otologic surgery [1-4]. Less commonly, the osseous defect is congenital or follows skull fracture, tumour or irradiation [2]. Nonetheless, since the introduction of more sensitive diagnostic tools in the last few years, reports of spontaneous or idiopathic brain herniations have increased [2].

The most common clinical presentation of encephalomeningocele is a conductive or mixed hearing loss or otitis media with effusion [2,4], other highly suspicious clinical presentations of spontaneous brain herniation and more specific symptoms [1,4] includes intracranial infection like meningitis and brain abscess, cerebrospinal fluid leak, epilepsy and aphasia [4], which are occasionally sentinel events [2]. The primary modality of treatment is surgery [2], and the management strategy of meningoencephalic herniation may differ depending upon the etiology, position and size of the bony defect, active cerebrospinal fluid leak, preoperative audiometry and the presence of active infection [1,4]. Whereas the Transmastoid Approach (TM) is generally useful for single, small (<1cm²) or medium-sized (1 to 2cm²) dehiscences [2,4] located at the level of the tegmen mastoideum or antri when the ossicular chain is not involved, a Middle Cranial Fossa Approach (MCF) is preferred for large or anteriorly located defects or when active infection is present [1,2,4]. The advantage of MCF is the possibility to reach bony defects located anteriorly without any manipulation of the ossicular chain [1].

Case Report

MVPM, 36 years old, female. Patient complaining of ear pain, tinnitus pulsatile and hearing loss for 1 year. She didn't have a history of trauma, high BMI or hydrocephalus. She consulted otolaryngologists several times and received treatment for otitis media without success. Physical examination showed right tympanic membrane hyperemia in the upper quadrant, without other alterations. Audiometry was performed evidencing conductive hearing loss in the left ear with a 20db gap. Magnetic resonance of 8^o nervous evidencing formation with sign similar to the cerebral parenchyma in T1, T2 and FLAIR and contours rounded occupies part of the epitympanum and the mastoid antrum to the right, measuring about 0.8 x 0.7 x 0.6 cm, underlying bone failure in the tympanic tegmen. Such findings allow us to consider the possibility of meningoencephalocele. Reduction of encephalomeningocele via mastoid was performed the patient had a very satisfactory evolution after surgery and no more complaints (Figure 1).



Figure 1: Pre operative , the green arrow shows the encephalomeningocele.

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