

Fungus Cerebri - A Case Report

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Abstract

Fungus cerebri is an extremely rare disease. The etiopathogenesis attributed to the condition are long standing mastoiditis, temporal lobe fracture and the most important common cause is iatrogenic (post-operative to mastoidectomy). The clinical diagnosis is the main stay which is supplemented by imaging studies. The most common herniated part of the brain is the temporal lobe, but cerebellar herniations are also seen. Surgery is the main stay in the treatment of brain herniation into the mastoid or middle ear cavity which can be neurosurgical, otosurgical and combined approaches. In this case report, we will be discussing a case of fungus cerebri.

Keywords: Fungus Cerebri; Meningoencephalocele; Brain Fungus; Mastoidectomy

Introduction

Fungus cerebri means herniation of brain and dura outside the cranial cavity into the mastoid cavity and middle ear. It is an extremely rare complication. Earlier it used to be seen in cases where a transmastoid approach was taken to drain a brain abscess complicating mastoiditis. The intense fear of this complication was an important factor in improvising the drainage method. It was done through a clean, separate burr hole instead off via a mastoid cavity [1]. The herniation of brain into the temporal bone was first described in 1902 by Caboche in the French Literature. Since 1902 many terms such as brain hernia, brain fungus, prolapse of brain, cerebral hernia, fungus cerebri, meningoencephalocele or encephalocele have evolved to describe this pathology [2]. Nowadays due to the surgical expertise, there is a decline in its incidence. This rarity has now caused a delay in the diagnosis. This delay results in the development of serious complications such as cerebrospinal fluid leak, meningitis, epidural or subdural abscess. Meningoencephalocele and encephalocele are encountered in less than 1% of the revision mastoid surgeries [3]. The predisposing factors for fungus cerebri are prior ear surgery (mastoidectomy),

chronic meningitis, cholesteatoma and active chronic otitis media. The most common sites of herniation are tegmen tympani and tegmen antri [4]. It develops as a sequel to defects in dura and skull. The brain tissue herniates through the tegmen defect into the mastoid cavity. This herniation may or may not be accompanied by cerebrospinal fluid leak. The ultimate management of fungus cerebri consists of wide surgical exposure, excision of the necrotic and herniated brain tissue, watertight intact dural closure and layered closure of the overlying soft tissues along with the correction of the underlying pathology of the increased intracranial pressure [1].

Case Report

A 54-year-old male who was apparently asymptomatic until 15 years of age, developed scanty, foul smelling right ear discharge which was insidious in onset and was gradually progressing with no specific aggravating and relieving factors. Subsequently he had an RTA with a head injury and ear bleed. During this time, he was diagnosed to have right atticofurrow disease and he underwent Right Tympanomastoidectomy. Patient was asymptomatic post-

surgery. 2 years later he developed watery discharge from the right ear with ear block. He was diagnosed to have right aural polyp in his native place and was treated with medications and ear drops. The patient was persisting to have watery ear discharge without any improvement and also subsequently developed headache. He also had severe tinnitus in the right ear. There were no symptoms of giddiness. At this point of time patient was referred to us through the surgeon who operated previously.

On examination, the left ear was normal. The right ear had a huge mass which was pinkish in colour, pulsatile, soft and insensitive mass which did not bleed on touch (Figure 1). The mass was completely filling the external auditory canal and was attached to its skin. Radiological examination revealed a heterogeneously enhancing mass filling the right EAC, middle ear and mastoid cavity with destructive erosion of tegmen tympani. Middle ear ossicles seemed eroded and could not be visualised. There was definitive defect in the tegmen tympani through which the brain tissue was seen herniating. Therefore, the diagnosis of Fungus cerebri was

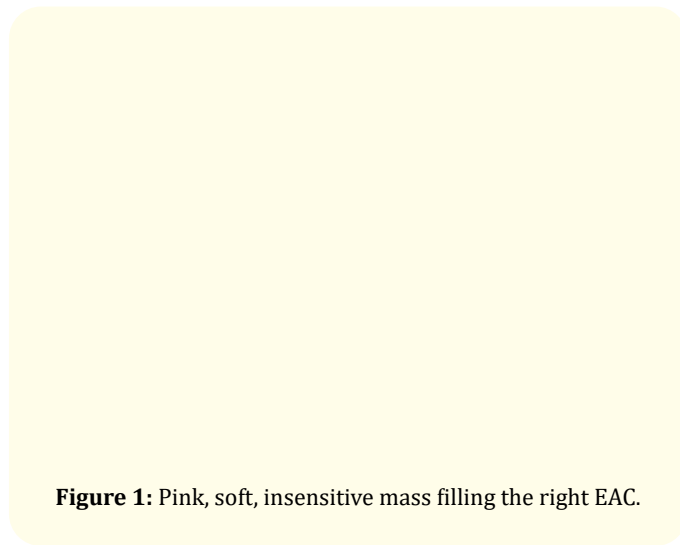


Figure 1: Pink, soft, insensitive mass filling the right EAC.

confirmed.

The Patient was then taken up for surgery after getting a proper written, informed consent. The surgery was done under general anesthesia along with a neurosurgeon. A transmastoid postaural approach was done and temporal bone chip was drilled out. Meningoencephalocele was seen herniating through the defect (10 x 6 mm) in tegmen tympani. The brain tissue was exposed after

carefully incising the dura. The herniated part of brain was then cauterized and removed. Margins of the defect were freshened and a bone grafting was done to seal the defect. Intra aurally the defect was closed using fascia lata and temporalis muscle flap which were all glued using Tissue Glue. The facial ridge was then lowered and the cavity was smoothed. The temporal bone chip was then replaced and secured using screws and plates. The wound was then closed in layers. Patient tolerated the procedure well with no intra operative nor post-operative complications. The histopathological examination confirmed the tissue as glial tissue.

Discussion

There are not many case reports of meningocele or meningoencephalocele herniation into the mastoid cavity or the middle ear cavity. There is one case of fungus cerebri reported by Saurabh Varshney, et.al in the Indian Journal of Otolaryngology in 2013 [12]. The common etiological factors include Chronic otomastoiditis, congenital dehiscence, skull base fractures and the most common is iatrogenic (post mastoidectomy surgery). Spiros Manolidis, in his study stated that the predisposing factors for herniation of dura into the middle ear and mastoid are prior ear surgery (radical or modified radical mastoidectomy), meningitis, cholesteatoma and active chronic otitis media [2,4]. Iatrogenic injuries incurred during mastoidectomy accounts for more than 59 to 77% of the cases of fungus cerebri [2,5]. The defects in the dura occur due to various reasons such as spontaneous, congenital and idiopathic [6], traumatic [7], iatrogenic and inflammatory [8] pathologies.

The tegmen plate is the commonest site of dural defect [4]. The dural herniations can be classified as pedunculated or sessile. Patients presenting with dural herniations also commonly have associated infections, CSF leak, and neurological complications. The commonly associated otological symptoms are hearing loss, tinnitus and trapped squamous epithelium. The most commonly herniated part of the brain is the temporal lobe, but cerebellar herniations are also seen. The case literatures reported, state that if the arachnoid remains intact, a meningoencephalocele occurs associated with the potential of a CSF leak into the ear and recurrent meningitis. An encephalocele without a CSF leak is seen in cases where the arachnoid matter is not intact.

The diagnosis is mainly based on a high index of suspicion, proper history taking, clinical examination and radiological evalu-

ation. High resolution computed tomography and Magnetic resonance imaging play a significant role in confirming the diagnosis and also to assess the extent of lesion [5]. Dural herniation into a post-operative mastoidectomy cavity is extremely rare condition which could radiologically be confused with a recurrent cholesteatoma on a CT scan.

Therefore, MRI is absolutely mandatory for appropriate diagnosis by differentiating cholesteatoma and inflammatory tissue from brain herniation.

Surgery is the primary and definitive treatment. There are numerous techniques used for the closure of dural defect which can be neurological, otological or combined approaches [6]. The otological approach is mainly transmastoid and is done in very small defects. The neurosurgical approach is through the middle cranial fossa or temporoparietal craniotomy. Combined approaches are advocated in very large herniations [9,10]. The herniated brain tissue is excised after cauterization as it is nonfunctional [5,11]. Various materials can be used to seal the defect in the tegmen plate like temporalis fascia, conchal cartilage, titanium mesh, cadaveric dura, muscle, bone, proplast, fibrin glue, etc [2].

Conclusion

Fungus cerebri is commonly seen as a complication of mastoid surgery. It can be prevented if appropriate care is taken while using burrs or diathermy so as to not injure the dura. A high index of clinical suspicion with meticulous examination is required to diagnose this condition as it is extremely dangerous. A vast clinical experience is absolutely mandatory so as to arrive at an early proper diagnosis and to prevent manhandling of the mass. Surgical management with a good team of ENT, Neurosurgeon, Radiologists remains the mainstay treatment. Also, early diagnosis and intervention is the key note in preventing the impending complications.

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