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CASE REPORT

Idiopathic herniation of the cerebellum into the mastoid process with internal jugular vein thrombosis

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ABSTRACT

Spontaneous herniation of cerebellum into mastoid air cells is a rare occurrence and has not been reported in the recent literature. We report a case of 37-year old man presenting with right ear pulsatile tinnitus and intermittent imbalance for the last 12 years. CT temporal bones showed a benign looking destructive lesion of posterior aspect of the right petrous bone.

Keywords: Mastoid; Tinnitus; Cerebellum.

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INTRODUCTION

Spontaneous cerebellar herniation into pneumatic system of temporal bone has not been reported in literature. The presenting signs and symptoms in normal middle ear create a diagnostic dilemma to the clinicians; neuroimaging studies have become an essential tool for diagnosis and treatment planning. Herniation of the cerebellum into mastoid bone could be congenital or acquired, and the later could be traumatic or spontaneous. Herniation may be a meningocele, meningoencephalocele or encephalocele. A large defect in the posterior wall of the petrous bone herniates the cerebellum into the mastoid and middle ear. This case report presents a case of spontaneous cerebellar herniation into mastoid system, without involvement of the middle ear.

CASE REPORT

A 37-year old man, working in Saudi Arabia presented to the ENT OPD with complains of tinnitus, intermittent imbalance, decreased hearing and reduced coordination on the 16th September 2019. He reported having tinnitus in the right ear and intermittent imbalance for the last 12 years. Recently, he noticed a significant hearing loss in right ear over the previous 6 months, along with some reduced coordination in the right side, for a few weeks; so much so, he had to shave with his non-dominant left hand. There was no significant past history except for head trauma in childhood, aged 2 years.

On examination, no neuro-otological signs were present, baring the finger-nose-test on the right, which, revealed poor coordination on his right side. Otoscopic examination resulted in a normal tympanic membrane bilaterally. Pure tone audiometry showed a unilateral 60db right sensory neural hearing loss.

A temporal bone Computed Tomography (CT) was done which demonstrated a destructive lesion in the posterior aspect of right petrous bone, continuous with posterior fossa between the internal auditory meatus and jugular foramen, which was diagnosed radiologically as a cholesteatoma or cholesterol granuloma. Mastoid process was fully occupied by mass with thrombosis of internal jugular vein [Figure 1(a-b)]. The findings on CT and the progressive clinical features warranted surgical exploration.

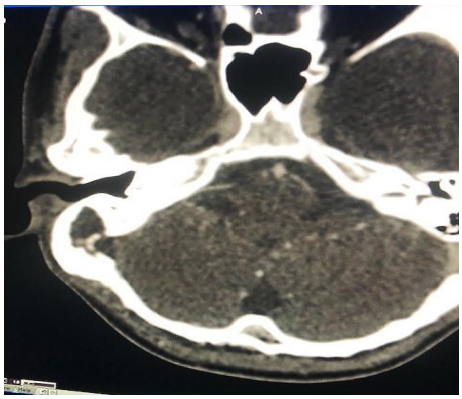

Figure 1(a) Herniation into Mastoid Process

Figure 1(b) Thrombosis of Internal Jugular Vein (arrow)

During the procedure [Figure 2(a-b)], a right post auricular incision extending to the upper part of the neck was done and mastoid exploration showed herniation of cerebellum into mastoid process with thrombosis of sigmoid sinus up to the upper part of external jugular vein. The necrotic brain tissue with thrombosed part of the sigmoid sinus along with the internal jugular vein was removed with ligation of the internal jugular vein.

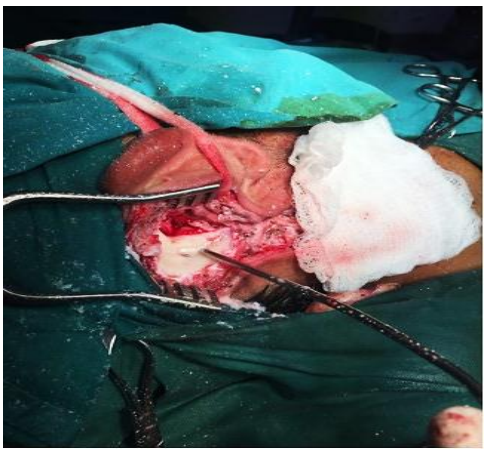

Figure 2(a): Peri-operative Findings

Figure 2(b): Postoperative right mastoid and neck exploration

The defect was closed with temporalis fascia, rotational temporalis muscle flap and fibrin glue. The wound was closed in two layers and recovery was uneventful with no CSF leakage and was discharged home on 18th September 2019. Follow up appointment were arranged after three and six months.

DISCUSSION

Spontaneous herniation between temporal lobe and middle ear or mastoid has been reported, however, spontaneous herniation of the cerebellum into the mastoid air is relatively rare. In this patient, the history of trauma at 2 years of age could have been a predisposing factor for the herniation.

Factors that predispose to herniation of brain tissue include infection, high intracranial pressure and raised cerebral volume.[1-3] The herniated tissue is non-functional and can be safely excised without neurological sequelae and excision of herniated brain tissue is the accepted treatment.[4,5]

The incidence of the tegmen defect have been reported to lie between 20% to 34% from 1965 to 1986.[6-8] Intact dura resists both infection and mechanical stress and the shearing forces between the two elastic layers of dura over the defective support may lead to herniation of brain tissue. In 1960, Schurr described a cerebellar herniation of posterior surface of temporal bone as an extremely rare occurrence.[9] Posterior fossa dura defects secondary to arachnoid granulations have been observed more recently. Current theory states that the defect develops from erosion of the granulation in the bone surrounding sigmoid sinus and Gacek postulated that only arachnoid granulation larger than 3.5mm were associated with bony erosion.[10]

In this case, reduced coordination on the right side, the inability to shave with right hand and poor finger nose test, showed progressive cerebellar damage. Pre-operative CT and MRI are useful tools to assess the extent of bone defect and brain tissue damage when available. The trans-mastoid approach was used for this patient, other approaches that could be used are transcalvarial and trans-temporal.

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