

Petrous bone epidermoid cyst caused by penetrating injury to the external ear: Case report and review of literature

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ABSTRACT

Epidermoid cysts are histologically benign, slow-growing congenital neoplasms of the central nervous system that may arise from retained ectodermal implants. The epidermoid lesions are generally caused during the 3rd to 5th week of gestation by an incomplete cleavage of the neural tissue from the cutaneous ectoderm, though it can also happen later in life due to introduction of skin elements by skin puncture, trauma or surgery. We present this unique case of a petromastoid epidermoid cyst associated with ipsilateral cerebellar abscesses, presenting 20 years after a penetrating trauma to the external auditory canal. Radical excision of both lesions and revision of the previous fistulous tract was performed. We present the diagnostic challenge and the operative treatment of this unique case, which to our knowledge is the first where an epidermoid cyst and an adjacent brain abscess occurred as a result of a single traumatic event.

Key words: Epidermoid cyst, penetrating trauma, petrous bone, surgical treatment

Introduction

Epidermoids are slow-growing congenital lesions of ectodermal origin, representing approximately 1% of all primary intracranial tumors.^[1-4] The cerebellopontine angle (CPA) is one of the most common sites affected,^[5,6] wherein the patients generally present with symptoms of cranial nerve, cerebellar or brainstem dysfunction, along with possible hydrocephalus and meningeal irritation.^[1,5-9] Hyperactive dysfunction of cranial nerves, such as trigeminal neuralgia and hemifacial spasm, has also been reported in association with this rare tumor.^[10] Microscopically, the capsule of the epidermoid cyst consists of a layer of avascular stratified squamous epithelium. The contents are mainly composed of keratin in concentric layers and cholesterol in a solid crystalline state. Patients with epidermoid cyst occasionally

experience complications such as aseptic meningitis and delayed postoperative hemorrhage. Intracranial epidermoids is considered to be congenital in etiology, though trauma is known to cause epidermoids in spine and elsewhere. In this report, we present a unique case of a patient presenting with cerebellar dysfunction, found to have a petromastoid epidermoid cyst in association with an ipsilateral cerebellar abscesses, following a penetrating trauma sustained to the external auditory canal canal about 20 years ago.

Case Report

History

This 54-year-old logger presented with symptoms of incoordination and underwent a computed tomography (CT) scan in August of 2010, which revealed an extra axial mass in the posterior fossa located in the mastoid and the sigmoid sinus region. He had a clinical follow-up visit by an otologist-ENT physician (SS) in December 2010, during which a fistulous tract from the external auditory canal was noted and surgical treatment was recommended, but delayed due to socioeconomic problems. The patient provided the history that he had sustained a penetrative trauma to the external ear canal by a small wooden stick when he was logging about 20 years ago, which was removed from the ear with uneventful healing.

Over the next 2 weeks, he noticed increasing blurry vision, tinnitus, problems with balance, and sleepiness. He was transferred to our hospital for treatment.

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Neurological examination and laboratory

On examination, the patient was alert with normal cranial nerve examination and afebrile. He had mild dysmetria of the right upper extremity. Preoperative blood, urinal and serological examinations were normal; no immunosuppression was detected; lungs were clear, and no skin infections were present.

Radiological findings

A cranial CT scan [Figure 1] revealed a hyperdense extraaxial mass in the posterior fossa located in the sigmoid sinus and mastoidal regions. There was significant erosion of the mastoidal bone. There was also edema in the adjoining cerebellar hemisphere. Magnetic resonance (MR) imaging [Figure 2] and magnetic resonance venogram (MRV) imaging revealed a space-occupying lesion filling the right mastoid air cells (32 mm AP x 27 mm transverse x 32 mm craniocaudal), that was heterogeneously hyperintense on T2, mildly heterogeneously hyperintense on T1 and showing restricted diffusion. The lesion showed minimal enhancement. There were two adjoining ring-enhancing lesions in the right cerebellum, which had cystic central area with layering debris. There was surrounding T2 hyperintense vasogenic edema in the right cerebellum and mass effect on the fourth ventricle and right side of the brainstem. The right sigmoid sinus appeared to be occluded.

Surgery

The patient underwent surgery via a temporal and retrosigmoid skin incision. A very large vascularized flap consisting of the posterior half of the temporalis muscle and pericranium was elevated and left *in situ* for subsequent repair. A retrosigmoid craniotomy was performed. The mastoid cortex, which was very thin, was removed and exposed. This quickly gave way to what appeared to be an epidermoid cyst [Figure 3]. It had a typical pearly white capsule with dirty white and pearly white contents, replacing the mastoid and posterior petrous bone. The dura mater was intact but was associated with granulation tissue. The capsule was carefully resected from the mastoidal area and the petrous bone, and was gently peeled away from the facial nerve. The facial nerve was left *in situ* and stimulating at 0.1 mA. The fistulous communication between the external ear canal and posterior fossa was clearly demonstrable. The aditus ad antrum was also displayed. The cyst and the granulation tissue were seen occluding the sigmoid sinus. The cerebellar dura was opened just adjacent to the epidermoid cyst in the location where the sigmoid sinus would have been. The cerebellum was adherent to the dura mater laterally, and at a depth of 0.5 cm, a cerebellar abscess with yellowish pus was seen, which was contiguous to a smaller abscess cavity. Complete resection of abscess and its capsule was achieved. Following this, the dura mater was closed in a watertight fashion [Figure 4a]. The vascularized temporalis-pericranial flap was rotated to cover the fistula over

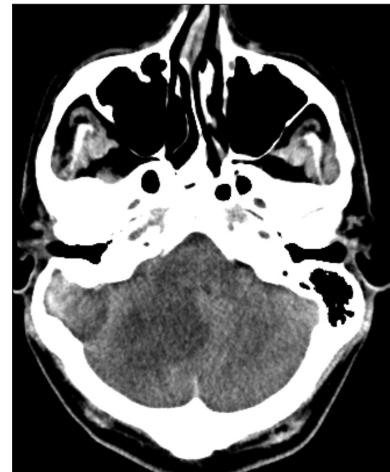


Figure 1: CT scan of the brain without contrast demonstrating a well-defined hyperdense mass occupying the right petromastoid region. Adjacent cerebellar edema compressing the IV ventricle is also shown

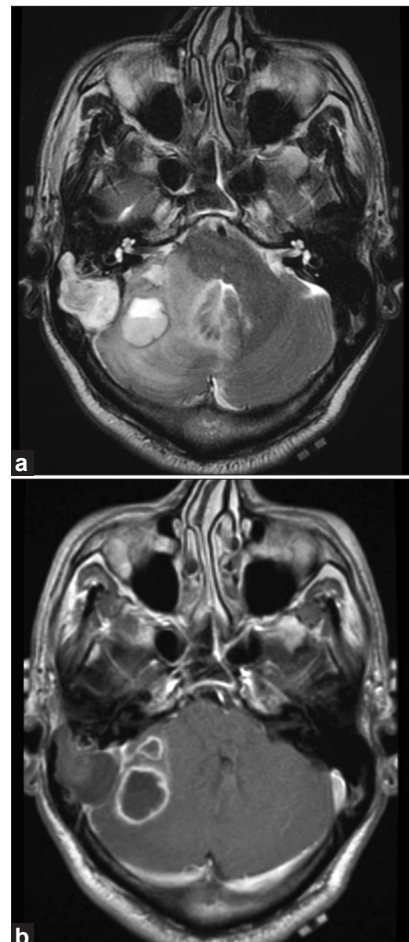


Figure 2: (a) Axial T2-weighted MR imaging demonstrating space-occupying lesion filling the right mastoid air cells that is heterogeneously hyperintense and associated with vasogenic hyperintense edema in the right cerebellum; (b) T1-weighted MR imaging with gadolinium contrast enhancing demonstrating no contrast enhancement of the mastoid lesion and two ring enhancement lesions (abscesses) in the right cerebellum with mass effect on the fourth ventricle and right side of the brainstem

the external ear canal as well as the middle ear and secured with some sutures [Figure 4b]. No bony reconstruction was done because of the infection.



Figure 3: (a) Intraoperative view of the epidermoid cyst in the mastoid region; (b) Gross view of the epidermoid cyst after radical resection

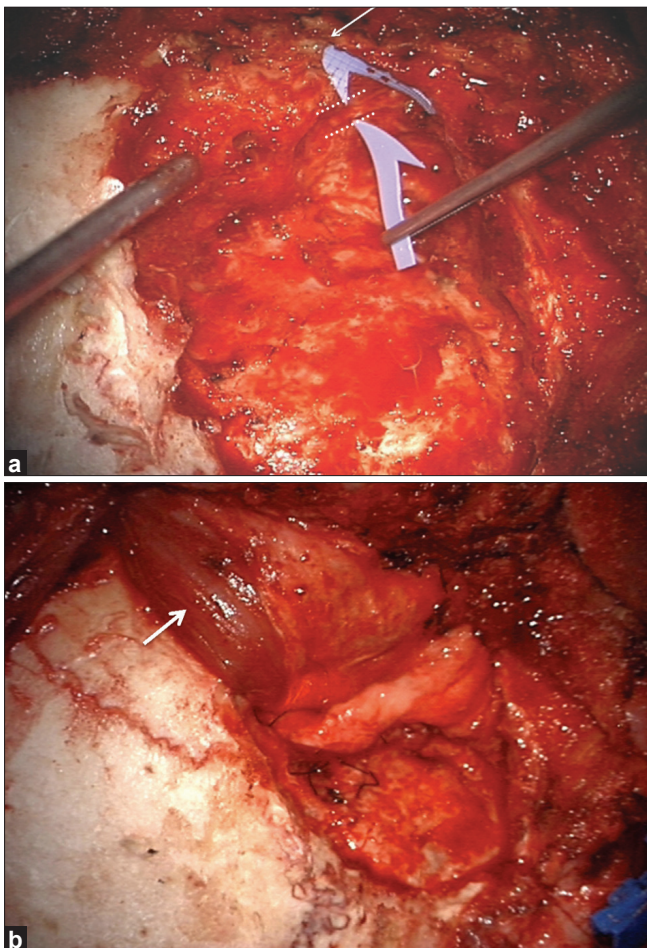


Figure 4: (a) The superior arrow indicates the opening of the fistulous tract (arrow) into the mastoidal area, and the facial nerve is shown between dotted lines; (b) Intraoperative image of the vascularized temporalis flap to cover the fistula over the external auditory canal as well as the middle ear (arrow)

Histology and microbiological findings

Histology revealed typical microscopical features of an epidermoid cyst [Figure 5] such as keratinized stratified epithelial lining of the cyst wall and central cholesterol and keratin debris. Cultures of the abscesses pus demonstrated the presence of *Serratia Marcesens* and coagulase negative Staphylococcus.

Outcome and follow-up

The patient did well after surgery. A postoperative MR imaging (not shown) of the brain, obtained 24 hours after surgery, demonstrated complete excision of both lesions without postoperative complications. The patient underwent intravenous antibiotic therapy (Meropenem and Vancomycin) for 6 weeks; and two months after surgery, he was doing well, and MR imaging [Figure 6] showed complete resolution of infection and the epidermoid cyst.

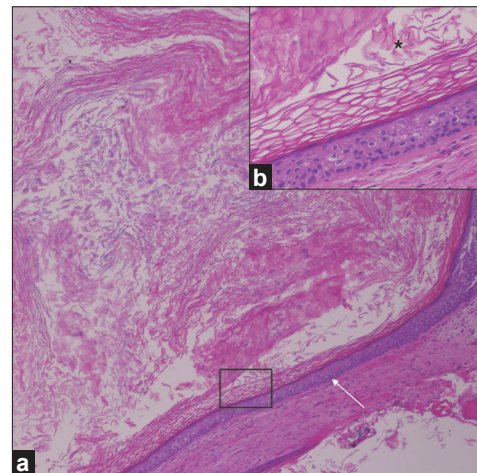


Figure 5: (a) Low power (40x) (a) and high power (200x) (b) H and E staining images demonstrating the characteristic keratinized stratified squamous epithelium lining (arrow) of the epidermoid cyst along with a superficial multilayer keratinaceous surface and keratin debris. In 5b the details of the anucleated cell ghosts (*) is seen better appreciated

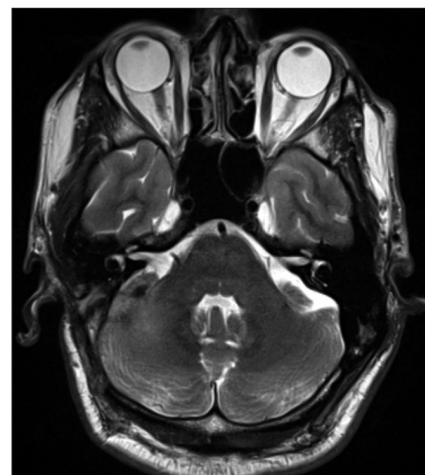


Figure 6: T1-weighted, gadolinium enhanced MR imaging axial view, obtained six weeks after surgery showing complete radical excision of the lesions, and healing after antibiotic therapy

Discussion

Epidermoid cysts or tumors were first described by Cruveilhier^[11] and designated the most beautiful tumors of the body by Dandy.^[12] Although the formation of epidermoid lesions is caused during the 3rd to 5th week of gestation by incomplete cleavage of the neural tissue from the cutaneous ectoderm, the mechanical introduction of such skin elements can also occur later in life by any mode of skin puncture.^[13] Epidermoid cysts have been reported as sequelae of trauma and surgery in bone, cartilage, and abdominal organs.^[14,15] The formation of epidermoid cysts after lumbar puncture has also been documented, most commonly in neonates, although at least three reports document their occurrence in adult patients.^[16,17] In the literature, three previous reports indicated the intracranial development of epidermoid cysts after head trauma or surgery.^[18-20] Epidermoid cysts have a thin capsule of stratified, keratinized squamous epithelium.^[21] They grow linearly as a result of desquamation of epithelial cells, which later break down into keratin a cholesterol. However, they may also expand and become enormous by neoplastic cellular growth. Estimates of the incidence of epidermoid cysts have ranged from 0% to 8% of all expansive intracranial lesions.^[1,21-23] The cisterns of the CPA and parasellar region are the most common site for development of an epidermoid cyst.^[1,6,22,24-29] On CT scanning, these lesions generally appear as well-defined lobulated hypodense masses, and occasionally display calcification in the cyst wall. On MR imaging, epidermoid cysts typically show low signal intensity on T1-weighted images and high intensity on T2-weighted images. Diffusion-weighted (DW) MR imaging has been found to be helpful in the differential diagnosis from arachnoid cysts, inflammatory cystic lesions, and dermoids.

The symptomatic onset in epidermoid cysts is usually slow, lasting 2 or more years, with headache as the most common symptom. Although, some patients with remitting signs and symptoms^[27] or with rapid onset^[6,25] have been reported. Epidermoid cysts of the CPA cause the symptoms and signs of a slowly expanding mass in that region,^[30] including ataxia, nystagmus, facial pain, paresthesias, and weakness.^[23,24,26,31-34]

Our case has unusual features such as: epithelial cell implantation into the cranium by penetrating injury, the location of the cyst, the concomitant presence of an adjacent cerebellar abscess, and the long delay to clinical presentation. Although, unusual CT and MRI appearance of intracranial epidermoid cysts has been described, hyperintensity in the CT without hemorrhage and the heterogeneously hyperintense appearance on both T1 and T2 weighted imaging and no contrast enhancement on both sequences seen in our case are interesting features. We speculate leakage of the irritant cyst content and resultant formation of granulation tissues inside and in the wall of the cyst as the cause for unusual radiological presentation. The penetrating trauma of the external auditory

canal 20 years ago as an underlying cause for both lesions makes this case unique.

In conclusion, implantation of skin causing epidermoid cyst is well known in the lumbar area, after repeated lumbar punctures with the use of stylet. Severe head and spinal trauma were postulated as a causative factor for the formation of intracranial epidermoid cysts. To our knowledge, this is the first documented case of an implantation induced epidermoid cyst and an adjoining brain abscess caused by a small penetrating trauma through the external auditory canal. We postulate that due to persistent fistula, there was secondary infection and cerebellar abscesses. The appropriate treatment consisted of complete removal of the epidermoid cyst and cerebellar abscesses followed by vascularized reconstruction of the infected site and the fistulous tract, which allowed healing and excellent outcome of the patient.

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