Volume : 3 | Issue : 2 | Feb 2014 • ISSN No 2277 - 8160

Research Paper | Medical Science

Epidrmoid Cyst in Cerebellopontine Angle (Brain)- A Case Report

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ABSTRACT

Brain stem epidermoid cysts are extremely rare lesions. Management of the epidermoid cyst is decompression of cyst contents and removal of cyst capsule. But in some cases resection of the cyst may result in a poor outcome because of cyst wall adhesion into the brain(8) stem.Epidermoid cyst, or pearly tumor, is congenital in origin and accounts for about 1% of intracranial tumors(1). It is benign extra cerebral intradural lesion and in about 40% of cases is located in cerrbellopontine angle(9). Epidermoidtumor is consist of keratin with cholesterol in a solid crystalline state and avascular nature(10). The cyst content derived from desquamated epithelial cells composed of mainly keratin in concentric layers(11). Clinically epidermoid cysts are present with headaches followed by cranial nerve deficits, cerebellar symptoms and raised intra cranial pressure. Other cystic lesions in brain can be cystic schwannoma, cystic meningioma, hemangioblastoma, pilocytic astrocytoma, glioblastoma, craniopharyngioma, dermoid cyst, arachnoid cyst, and inflammatory cystic lesions are nerocystocercosis, toxoplasmosis, paragonimiasis.

Epidermoidtumor shows calcification in a minority of cases, and rarely an epidermoid cyst may be hyperdense due to haemorrhage, saponification or high protein content.

KEYWORDS : cerebellopontine angle, cystic lesions in brain

INTRODUCTION

Inttracranial epidermoid cysts are congenital lesions which account for about 1% of all intracranial tumours[2,3]. They result from inclusion of ectodermal elements during neural tube closure.

Although predominantly congenital, epidermoid cysts are usually very slow growing and as such take many years to present. Typically patients are between 20 and 40 years of age[6]. There may be increased prevalence in males, although this is not found in all series[5-6].

An uncommon association exists with anorectal anomalies, sacral anomalies and pre-sacral mass, and is known as the Currarino triad[5].

Signs and symptoms of epidermoid cysts are due to gradual mass effect, with presentation including[4] headaches - most common, cranial nerve deficits, cerebellar symptoms, seizures, and raised intracranial presure. Recurrent aseptic meningitis is uncommon but recognised, similar to the less common dermoidcyst[4].

Epidermoid cysts may be congenital (most common) or acquired (post surgical or post traumatic implantation). Pathologically, intracranial epidermoid cysts are identical to the petrous apex and middle earcongenital cholesteatomas. They differ from dermoid cysts which have both epidermal and skin appendages such as hair and sebaceous cysts and mature teratomas which have all three layers.

They have a thin capsule which is made of a thin layer of squamous epithelium, which macroscopically is white and pearly, and may be smooth, lobulated or nodular. Internal cystic components are often filled with desquamated epithelial keratin and cholesterol crystals; grossly appearing as a creamy, waxy material. It should be noted that some authors contest that epidermoid cysts are along a spectrum which includes cystic (adamantinomatous) craniopharyngiomas, which have some histological similarities[7]. Cholesterol and keratin components account for their distinct imaging characteristics[2].

Most common location is intradural(90%) in which 40-50% cases of cerebellopontine angle area[2].

third most common cerebellopontine angle mass, after acoustic schwannomas and meningiomas, and accounts for approximately 5-10% of all tumours in this region[2,4]. Other intradural location of epidermoid cyst are, suprasellar cistern, fourth ventricle, middle cranial fossa, spinal (rare).

CASE REPORT

A 8-year-old female child reported headache for 1 year. She had experienced severe dizziness, vertigo, and unsteady gait during the preceding 3 months. She also reported double vision when she looked to right side and experienced a choking episode 1 month earlier. Neurologic examination showed partial ptosis and partial facial palsy on the right side, tongue deviation to the right, hypoesthesia of the left upper trunk, and tandem gait impairment. There was no history of trauma.

MR imaging of the brain revealed an extra-axial, lobulated soft-tissue mass, measuring 48 × 30 × 36 mm, in the right cerebellopontine angle and prepontine cistern. Mild pressure erosion of adjacent petrous bone and partial encasement of the basilar artery were noted. Most of the lesion had homogeneous hyperintensity on both T1-weighted and T2-weighted images.

Grossly, specimen received consistent of multiple, grey-white, soft, flatten, membranous tissue along with dirty, creamy waxy material. All together measuring 4.8x3.5x3 cm. Taking multiple bits from tumor tissue for histological examination (eosin and hematoxylin)

Microscopically, the wall of the epidermoid cyst consists of a layer of keratinizing squamous epithelium without vascularity(fig.1).

The cyst content is derived from desquamated epithelial cells composed mainly of keratin in concentric layers and cholesterol in a solid crystalline state (fig.2). Histologic diagnosis was compatible with an epidermoid cyst.

Discussion

An epidermoidtumor is a congenital lesion that arises from inclusion of ectodermal epithelial elements[12]. The cerebellopontine angle is the

most common site of occurrence of intracranial epidermoid tumors, accounting for 5% of tumors in this region[13]. Grossly, epidermoid tumors are typically well-defined lesions with an irregular nodular outer surface and a shiny "mother of pearl" appearance[14,15]. Microscopically, the wall of the epidermoid cyst consists of a layer of keratinizing squamous epithelium without vascularity(fig.1). The cyst content is derived from desquamated epithelial cells composed mainly of keratin in concentric layers and cholesterol in a solid crystalline state[12,15].

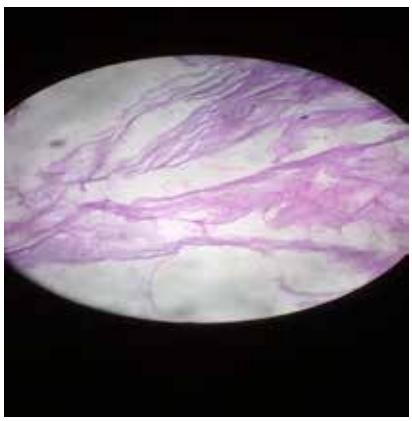


Fig. 1

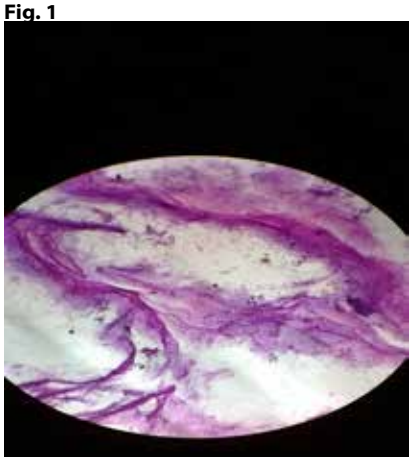


Fig. 2

Epidermoidtumors characterized surgically as being cystic and having a high lipid content comprising mixed triglycerides containing unsaturated fatty acid residues, and no cholesterol[11].

The differential diagnosis of cystic mass in the cerebellopontine angle can be cystic schwannoma, cystic meningioma,and arachnoid cyst, dermoid cyst.

A purely cystic schwannoma is very rare and can have an appearance similar to epidermoidtumor on MR imaging images. Findings of extension into the internal auditory canal and strong enhancement of the cyst wall may help to diagnose schwannoma.

Dermoid cyst, by contrast, are endowed with skin appendages including pilosebaceous glands units, accrine and occasionally,apocrine glands[16].

Cystic meningioma, soft, gray white,oval to hemispheric mass, cut surface shows solid with small cystic areas. Histologically, Cystic meningioma is named for its content of variably intercellular vacuoles. Cells are oval to spinle shapes with mild pleomorphism[16].

Arachnoid cysts are smoothly marginated, cause compression of adjacent structures, and are not enhanced after contrast administration[5]. Hemorrhage in any cerebropontine angle cystic lesion is not common and has rarely been reported in the literature.[15].

Epidermoid cysts typically show undulating margins and model their shape to conform to the cerebropontine angle. The cyst has a tendency to insinuate itself around the nerves and blood vessels in thecerebropontine angle.

In our case of an epidermoid tumor that showed variable signal intensity on T1 and T2 weighted MR images and histopathologically, diagnosis was compatible with an epidermoid cyst.

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