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Cerebellopontine angle epidermoid CYST: Case report

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Abstract
Epidermoid cysts are pearly tumors which are congenital in origin, benign, extra cerebral intradural lesions and about 40% of cases are located at cerebellopontine angle. A 27 year old female with complaints of headache, severe dizziness, vertigo, and unsteady gait since 6 months along with partial ptosis, partial facial palsy on the right side, tongue deviation to the right, hypoesthesia of the left upper trunk, and tandem gait impairment presented to the OPD of a Tertiary Care Hospital. Magnetic Resonance Imaging (MRI) was done which revealed a space occupying lesion on the right cerebellopontine angle along with enlargement of peripontine space. Lesion was removed surgically and was sent to histopathology department where it was diagnosed as epidermoid cyst.

Keywords: epidermoid cyst, cerebellopontine angle, CPA

Introduction
Intracranial epidermoid cysts are congenital and constitute about 1% of all the intracranial tumors [1, 2, 3]. They arise from ectopic inclusion of ectodermal cells during embryonic life while closure of the neural tube between the third and the fifth weeks intrauterine and are rare, slow-growing benign lesions [4, 5, 6]. On microscopic examination these usually show a cyst which is lined by stratified squamous epithelium, underneath seen is a central core of keratin flakes, desquamating cells and cholesterol [7]. Acoustic neuromas and Meningiomas are the most common tumors occurring at CPA, following which Epidermoid Cyst being the third lesion presenting at the same site [4, 6]. Other rare sites are parasellar region, petrous apex, chiasmal region, brainstem and intraventricular cavity [5]. Clinically patients present with slow growing mass compressing the cranial nerves, cerebellar and brainstem structures with usual age of presentation lying between the second and fifth decades of life [8]. Commonly patients with these tumors have a long history of tinnitus and hearing loss, occasionaly trigeminal neuralgia, facial paresis/ hemi facial spasm, headache, hydrocephalus and chemical / aseptic meningitis while vestibular symptoms are seldom seen [8].

Case Report
A 27-year-old female presented in the OPD with chief complains of headache, severe dizziness, vertigo, and unsteady gait since 6 months. She also reported diplopia while looking to the right side. Neurologic examination revealed 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. On radioimaging; MRI of the brain revealed a space occupying lesion on the right cerebellopontine angle with enlargement of peripontine space. (Fig -1)
On Gross Examination the received specimen consisted of multiple, grey-white, soft, dirty, creamy waxy material all of which aggregated to 5 cm.
On histology; sections stained with conventional Hemotoxylin and Eosin Stain showed a cyst wall lined by keratinized stratified squamous epithelium; cyst content being derived from desquamated epithelial cells comprising mainly of keratin in concentric layers and cholesterol in a solid crystalline state (Fig.-2, 3). Final histologic diagnosis was consistent with an Epidermoid Cyst.

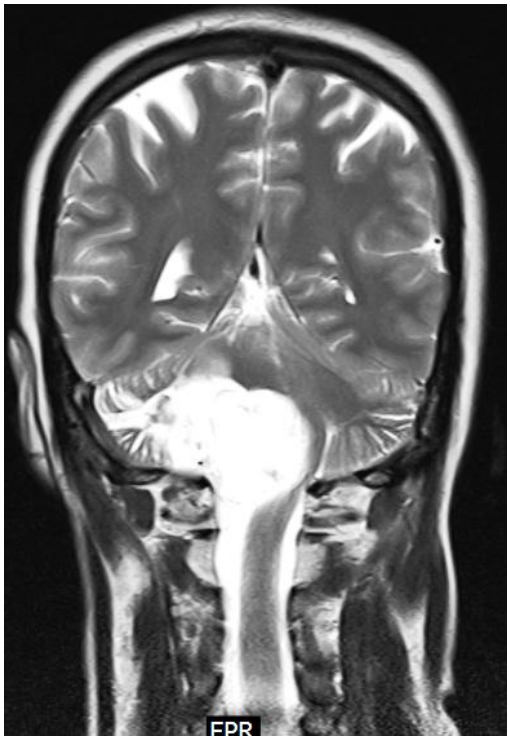

Fig 1: MRI shows high intensity lesion in the right CPA

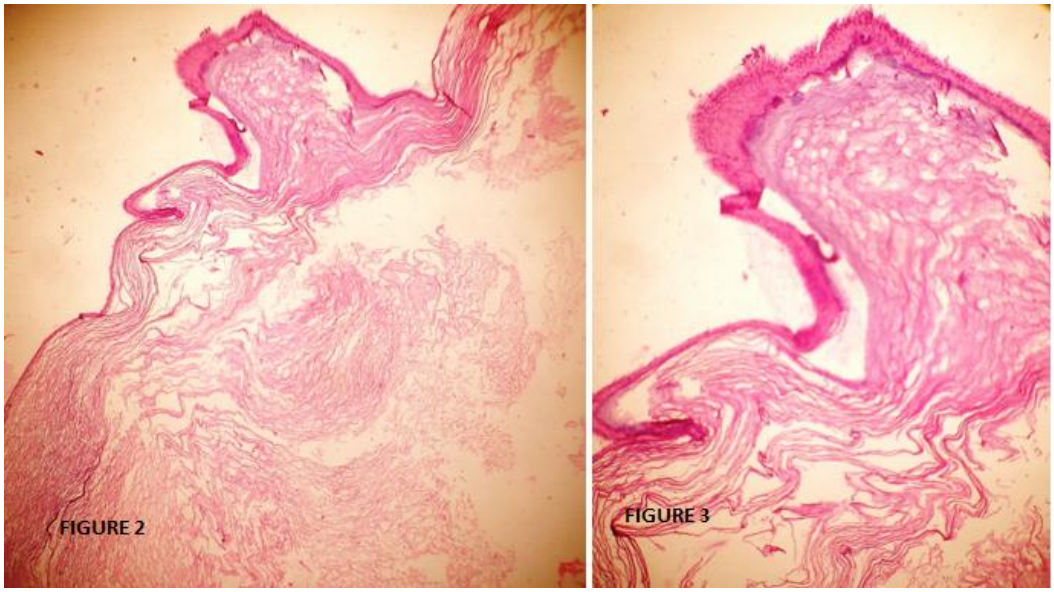

Fig 2 & 3: shows wall of epidermoid cyst lined by stratified squamous epithelium, underlying tissue shows concentric layer of lamellated keratin. (H&E 40X and 100X)

Discussion

Epidermoid tumor was first reported in 1683 by Duverney and their first epidermal origin description and their nomination as "epidermoid" was in 1854 by Remak [9, 10] an epidermoid tumor arises from inclusion of ectodermal epithelial elements and is congenital in origin. As mentioned earlier cerebellopontine angle is the most common site of its occurrence, accounting for 5% of tumors in this region [11, 12]. Epidermoid means "epiderm'' like suggesting that it's content arises mainly from the tissues which are normally present in the skin like keratinized stratified epithelium, keratin flakes or cholesterol and also interior of the cyst consists of "pearly white'', waxy tissue which is due to deposition of concentric lamellar keratin rich in cholesterol crystals, secreted by the basal cuboidal germinative stratus of the peripheral epidermal tissue [13, 14]. Signs and symptoms of epidermoid cysts are due to gradual mass effects which presents as headaches - most commonly, cranial nerve deficits, cerebellar symptoms, seizures, raised intracranial pressure and recurrent aseptic meningitis occurs uncommonly; similar to the less common dermoid cyst [15]. As in the case presented to us patient had similar complaints. Most common location of such tumors is intradural(90%) out of which 40-50% cases occur at cerebellopontine angle area, it is the third most common cerebellopontine angle mass, after acoustic schwannomas and meningiomas [15, 16] Thus reinstating the predlication of epidermoid tumor to the cerebellopontine angle as in the given case. Due to its slow growing nature it clinically manifests in later stages of life (35 years) and females are the most commonly affected gender [17]. Grossly, epidermoid tumors are typically well-defined lesions with an irregular nodular outer surface and a shiny "pearly white" appearance [15, 18]. Microscopically, sections studied reveal a cyst wall consisting of a layer of keratinizing squamous epithelium without vascularity. The cyst content is derived from desquamated epithelial cells composed mainly of Keratin in concentric layers and cholesterol in a solid crystalline state [11, 18] similar to our case.

Epidermoid tumors characterized surgically as being cystic and having a high lipid content comprising mixed triglycerides Containing unsaturated fatty acid residues, and no cholesterol [19]. A variety of cystic lesions other than epidermoid cyst occur at cerebellopontine angle such as cystic schwannoma, cystic meningioma, arachnoid cyst and dermoid cyst [20].

A purely cystic schwannoma is very rare and is very difficult to distinguish from epiderrmoid tumor on MRI due to its striking similarity with the former. If the lesion is extending into the internal auditory canal and there is strong enhancement of the cyst wall; it is diagnosed as schwannoma [20] Dermoid cyst, by contrast, are endowed with skin appendages including pilosebaceous glands units, eccrine and occasionally apocrine glands [20]. Cystic meningioma is characterized by soft, grey white, oval to hemispheric mass; on cut section it is solid with small cystic areas and histologically it shows variable intercellular vacuoles, cells are oval to spindle in shape with mild nuclear pleomorphism [20] whereas Arachnoid cysts have smooth margins, causing compression of adjacent structures and are not enhanced after contrast administration [21] Hemorrhage in any cerebellopontine angle cystic lesion is not common and has rarely been reported in the literature [18].

Conclusion

Thus to conclude, Epidermoid cyst is a rare congenital tumor of the central nervous system. The diagnosis of which require clinical history, complete neurological examination and radiological studies but the histopathological examination plays a pivotal role for the confirmation of same.

References

1. Eekhof JLA, Thomeer RTWM, Bots G. Epidermoid tumor in the lateral ventricle. SurgNeurol 1985; 23:189-92.
2. Boggan JE, Davis RL, Zorman G, Wilson CB. Intrasellarepidermoid cyst. J Neurosurg. 1983; 58:411-5.
3. Russel DS, Rubinstein U. Congenital tumors of mal-developmental origin. Edward Arnold(ed). Pathology ofTumours of the Nervous System, London. 1977; 24-64.
4. Son DW, Choi CH, Cha SH. Epidermoid Tumors in the Cerebellopontine Angle Presenting with Tri- geminal

Neuralgia. Journal of Korean Neurosurgical Society 2010; 47:271-77.
5. Chu CK, Tseng HM, Young YH. Clinical Presentation of Posterior Fossa Epidermoid Cysts. European Archives of Otorhinolaryngology, 2006; 263:548-51.
6. David EA, Chen JM, Posterior Fossa Epidermoid Cyst. Otology & Neurotology, 2003; 24:699-700.
7. Sigari A, Bellinzona M, Becker H, Samii M. Epidermoid Cysts of the Cerebellopontine Angle with Extension into the Middle and Anterior Cranial Fossae: Surgical Strategy and Review of the Literature ActaNeurochirurgica. 2007; 149:429-32.
8. Dutt SN, Mirza S, Chavda SV, Irving RM. Ra diologic Differentiation of Intracranial Epidermoids from Arachnoid Cysts, Otology &Neurotology 2002; 23:84-92.
9. Young Jeon JI, Min Kim J. Epidermoid Cyst of the Fourth Ventricle, Journal of the Korean Neurosurgery Society 2005; 38:478-80.
10. Remak R: Ein Beitrag zur Entwicklungsgeschichteder krebshaften Geschwulste. Dtsch Arch Klin Med 1854; 6: 170.
11. Gao PY, Osborn AG, Smirniotopoulos JG. Radiologic-pathologic correlation: epidermoidtumor of the cerebellopontine angle. AJNR Am J Neuroradiol 1992; 13:865-72.
12. Salazar J, Vaquero J, Saucedo G..Posterior fossa epidermoid cysts. Acta Neurichir 1987; 85:34–39
13. Robert H. Wilkins, Setti S. Neurosurgery Neuro-oncology: AnOverview, Epidermoi and Dermoid Tumors 2010; 6(1):655-661
14. Fornari M, Solero C L: Surgical treatment of intracranial dermoid and epidermoid cysts in children, Childs nervous system 1990; 6:66-70.
15. DeSouza CE, deSouza R, da Costa S. Cerebellopontine angle epidermoid cysts: a report on 30 cases. J. Neurol. Neurosurg. Psychiatr 1989; 52:986-90.
16. Chen CY, Wong JS, Hsieh SC. Intracranialepidermoid cyst with hemorrhage: MR imaging findings. AJNR Am J Neuroradiol. 2006; 27:427.
17. Winn R, Dacey R: Youmans Neurological Surgery, Epidermoid, Dermoid and Neurenteric Cysts. 2004; 5:1223-31.
18. Ochi M, Hayashi K, Hayashi T. Unusual CT and MR appearance of an epidermoidtumor of the cerebellopontine angle. AJNR Am J Neuroradiol 1998; 19:1113-15.
19. Horowitz B, Chari MV, James R. MR of intracranial epidermoidtumors: correlation of in vivo imaging with in vitro 13C spectroscopy. AJNR Am J Neuroradiol 1990; 11:299-302
20. Rosai and acherman's surgical pathology 2011; 9:1320-21.
21. Albright AL, Adelson PD, Pollack IF. Principles and practice of pediatric neurosurgery. Thieme Medical Pub 2007; 23:1411-19.