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An Enigma for treating doctors: Arachnoid cyst of Cerebello pontine angle (CPA) in an adult : A case report

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Abstract
Arachnoid cysts are benign developmental collections of cerebrospinal fluid (CSF). Arachnoid cysts are not very common more markedly arachnoid cysts of the cerebellopontine angle and are mostly asymptomatic. These arachnoid cysts comprise approximately 1% of all intracranial space occupying lesions [1]. Arachnoid cysts are considered developmental anomaly of the arachnoid membrane and arise from membrane duplication & splitting. Arachnoid cysts may also occur in other regions of the brain like suprasellar cistern or quadrigeminal cistern with middle cranial fossa being the most common site of occurrence with CPA being the next common site .Arachnoid cysts can expand & may cause signs & symptoms due to compression produced on the surrounding neural structures or increasing intracranial pressure. Arachnoid cysts at the CPA can enlarge enough to associate with hydrocephalus- suggesting that there is a direct relationship between the development of arachnoid cyst & abnormal CSF dynamics. Most commonly it is diagnosed by MRI scan with definitive investigations done for other specific complaints to rule out complications. Although, the definitive treatment for these arachnoid cysts is a retrosigmoid suboccipital craniotomy and microsurgical resection & fenestration of the cyst wall a wait and watch approach can also be done for if the subject does not have any complications as such after ruling out the other causes of complications if any.
Key words: *Arachnoid cysts, retrosigmoid , suboccipital, craniotomy*

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I. Introduction:

Arachnoid cysts are usually benign , cerebrospinal fluid filled sacs located in the brain or spinal cord & comprise approximately 1% of all intracranial space occupying lesions . However, lesions of the cerebello pontine angle account for 6-10% of all intracranial lesions.[2] Tumours of the CPA are mostly acoustic neurinomas and secondly, meningiomas – consist of approximately 85 to 90 percent and remaining lesions of CPA are primary cholesteatomas & facial nerve schwannomas. In this perspective, arachnoid cysts of CPA are rare in occurrence.[3] They are considered a developmental anomaly of the arachnoid membrane and arise from membrane splitting or duplication [4],[5]. Arachnoid cysts may also occur in other regions of the brain like suprasellar cistern or quadrigeminal cistern with middle cranial fossa being the most common site of occurrence with CPA being the next common site of occurrence. The arachnoid cysts enlarge enough to associate with hydrocephalus which points towards a direct relationship between the development of arachnoid cyst and abnormal CSF dynamics. So, arachnoid cyst can expand and may cause signs and symptoms by compressing surrounding neural structures by increasing intracranial pressure. Onset of symptoms is usually due to the compression of the brain , cranial nerves & obstruction of the CSF circulation. In case of the arachnoid cyst of CPA the major symptoms as reported by the few patients who had it mostly comprised of headache, diplopia, ataxia and 8th cranial nerve palsy. There maybe mirror movements of the extremities mostly of the upper limbs which maybe as a result of the arachnoid cyst compressing the brainstem & the pyramidal decussation. Mostly, the arachnoid cysts of CPA usually remains asymptomatic – so diagnosis is mainly by incidental finding during radiological evaluation for other reason. The onset of symptoms and signs is usually due to the compression of the brain, cranial nerves and obstruction of the CSF circulation. The major clinical symptoms of CPA arachnoid cysts are mainly ataxia & headache. Patients may also present with nausea, vomiting, cerebellar signs, character change, memory disturbance [6]. Other less common symptoms maybe mirror movements, diplopia. These cysts may also cause dysfunction of the particular cranial nerves like II, IV, VI(which causes diplopia). Dysfunction ov Vth cranial nerve may cause trigeminal neuralgia as well as it may lead to facial nerve dysfunction when it affects the VIIth nerve. Likewise when it affects theVIIIth nerve it may cause hearing loss, tinnitus, vertigo. When it affects the Xth nerve it results in hoarseness of voice & dysphagia [7] So, magnetic resonance imaging (MRI)of the brain usually points to the diagnosis with supporting tests like visually evoked potential(VEP), lumbar puncture for manometry and cytology forming some of the important tests to find out the cause of the complications produced secondarily due to the compressive nature of the arachnoid cyst of the

CPA. Surgical excision of the cysts by a retrosigmoidal suboccipital craniotomy approach or microsurgical resection and fenestration of the cyst walls is the definitive treatment of the arachnoid cysts of the CPA. For paediatric age group patients shunt independent surgical treatment as well as microsurgical fenestration and cystoperitoneal shunt through preauricular subtemporal keyhole craniotomy for the treatment of symptomatic patients can be done. A wait and watch approach is also advocated for asymptomatic patients as CPA arachnoid cysts are mostly incidental findings with cyst usually remaining silent. As the cyst contains osmotically active particles may get swollen at times and produce compressive symptoms. Mirror movements are involuntary movements , which occur in a muscle group or limb on one side of the body in response to an intentionally performed movement in corresponding contralateral muscle group or limb [6]. Mirror movements are of three types:

1) Physiologic form: This form usually presents at birth & disappears following maturation of the nervous system by the age of 8-10 years.
2) Hereditary form: Heterozygous mutations of DCC gene was found [7]
3) Pathologic form: It can be clinically isolated or associated with the nervous system disorders.

The disappearance of mirror movements generally occurs by the spontaneous disappearance of mirror movements by the end of the first decade if it is physiologic with family history excluded to rule out hereditary causes. There was no symptoms to suggest of a pathologic type of mirror movements. Arachnoid cysts contain cerebrospinal fluid and are benign cystic lesions that develop in the intra arachnoid space. Although the pathogenesis of those cysts are unknown- they are thought to be congenital ; Other causes of arachnoid cysts are infection, trauma, splitting abnormalities of the arachnoid membrane, alteration of the CSF flow and/ or change of the CSF pressure. In this article, a 26 year old girl with an arachnoid cyst of the left cerebello pontine angle that led to headache, diplopia and progressive dimness of vision –her case is an isolated case in an adult- so discussing the case.

II. Case Report :

A 26 year old girl presented with headache which was throbbing in nature of mild intensity mainly in parieto-occipital area off and on since a year with complaints of diplopia since one month alongwith progressive dimness of vision bilaterally since seven days. No previous history of trauma sustained to the head or no family history of any intracranial lesion. There was no history of vestibular symptoms, tinnitus, nausea, vomiting, meningitis. The otolaryngologic, neurologic and vestibular findings were within normal limits. On visually evoked potential (VEP) test- pattern shift VEP study reveals prolongation of P-100 latency (R>L) with normal amplitudes in both eyes which is consistent with bilateral demyelinating type visual pathway disorder (R>L).On fundus examination the disc margins appears blurred and diplopia on binocular vision. The deep tendon reflexes were normal with plantar reflex flexor bilaterally with Romberg's sign is negative with a provisional diagnosis of hydrocephalus with right optic neuritis with the investigation of MRI brain & orbit with MR venography, VEP both eyes. The MR venography of brain with the use of contrast was done & it revealed hypoplastic left transverse sinus seen with the other major sinuses showed normal course, calibre, luminal signal & drainage pattern. Rest of the superficial & deep veins otherwise show normal course, calibre, luminal signal and drainage pattern. Deep draining veins are normal including bilateral internal cerebral veins & venous angle with filling of the straight sinus with bilateral vein of Labbe very well visualised – so ultimately the impression of hypoplastic left transverse sinus with rest within normal limits. In the brain with contrast with the sequences used T2W, FLAIR, Diffusion, hemoaxial, T2W –coronal & T1W MPr3D- Sagittal with reconstruction & T2FS, T1FS, Obl.sagittal and coronal (for orbits) with post contrast T1FS-axial,coronal, sagittal with the findings of bilateral mildly prominent supratentorial ventricles seen more than the prevailing age. Well defined T2 hyperintense hypo on T1/ FLAIR cystic area noted at the left CP angle having no obvious contrast enhancement measuring 2.1 cm having no obvious diffusion restriction of a arachnoid cyst. The rest of the visualised part of both the orbits show normal globe with normal outline. Extraocular muscles are normal . No obvious evidence of their thickening , inflammation or edema seen . The optic nerves were normal in their course , caliber and signal pattern forming optic chiasma and optic tracts. No obvious thickening, edema or any mass lesion observed in the optic nerves. No obvious evidence of retinal detachment observed . Perorbital soft tissues are normal. No obvious intra or extraocular lesions seen. Rest of the visualised portions of the cerebral & cerebellar hemispheres with basal ganglia, thalamus, vermis & brainstem appears normal. Ventricular system & CSF spaces are normal with normal CP angles, optic nerve and optic chiasma and internal acoustic canal (IAC). No mass, infarct, hemorrhage, midline shift observed. Sella & parasellar regions are normal. Subsequently, the patient was planned for lumbar puncture and CSF manometry and cytological examination of the aspirated fluid to see for any raised intracranial pressure or infective pathology in the brain. The lumbar manometry as well as the cytologic examination of the lumbar fluid all the readings and counts came out within normal range . The MRI of the spinal cord was done with no significant abnormality detected . After all these investigative

procedures the young female was treated with steroidal injections to suppress the acute inflammatory episode brought by optic neuritis most probably and she responded to it with her ocular symptoms of headache, diplopia and blurring of vision improved. Arachnoid cyst of the CPA didnot produce any symptoms as such a wait and watch approach was adopted with successive follow up by MRI and consultancy with the onset of obstructive features . Repeated follow up consultation is to be done. So this arachnoid cyst of the cerebellopontine angle in case of this adult female didnot produce any compressive symptoms for which the outcome was probably better . But it needs followup as it may anytime enlarge and have the potency to cause compressive symptoms and may raise the intracranial pressure – producing emergency conditions. Presently no surgical interventions are required as such as arachnoid cysts are benign lesions but recurrence of the symptoms be it compressive / noncompressive cannot be denied for which regular follow up was advised.

III. Discussion:

The arachnoid cysts are most of the times asymptomatic. Compressive symptoms of the brain can occur following an increase in the osmotic gradient as there are osmotically active substances constituting the liquid content of the cyst. Another way of increase in the size of the cyst maybe formation of a valve mechanism between the arachnoid cyst & the subarachnoid space or there maybe secretion of liquid from the wall of the cyst ultimately leading to the enlargement of the cyst. Usually arachnoid cysts may become symptomatic by compressing the brainstem. The onset of the symptoms and elicitation of the subsequent signs followed by the investigative procedure was are entirely due to the demyelinating type visual pathway disorder with probably optic neuritis leading to the dimness of vision and headache plus diplopia which showed signs of improvement once the patient was treated with injectable methylprednisolone with the ocular symptoms of diplopia, progressive dimness of vision with headache remission. Symptoms as such was not produced by arachnoid cyst as such which didnot have the compressive symptoms of trigeminal neuralgia, vestibular symptoms like tinnitus , vertigo etc. Very few total about 30 cases are there with arachnoid cyst of CPA of which about 5 of them had compressive symptoms of trigeminal neuralgia even some may turn out to be quadrigeminal arachnoid cysts & not CPA arachnoid cysts.

IV. Conclusion:

CPA arachnoid cysts are rare clinical condition more so in an adult and the etiology and pathogenesis remains still an enigma for the medical fraternity as compressive symptoms produced by the arachnoid cysts may significantly put the treating doctor and the team of neurologist, neurosurgeon, otologist as well as the general practitioner at dismay regarding the diagnosis and which in turn may be the agony and decrease the QL as regards the young patient who can contribute significantly to the welfare of the society as such. Nowadays with the array of diagnostic modalities it is easy though to diagnose and allay the suffering of the patient and restore the quality of life of the patient to normalcy. The treatment based on the case to case approach as such as this is a very rare clinical condition with the use of operative procedures a still lesser entity with the risk and benefits of the surgical procedures if needed should be discussed at length with the patient & relatives for it to be successful. However, in most cases specially in adults without complaints and signs of compressive symptoms – a wait and watch policy can be safely applied with regular plan of follow up for at least upto a year after diagnosis.

Declaration of Conflict of interest :

It is declared that there is no competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References:

[1]. Dyck P, Gruskin P. Supratentorial arachnoid cysts in adults, Arch. Neurology 34 (1977), 276-9.

[2]. Brackmann DE, Arriga MA: Extraaxial neoplasms of the posterior fossa. Schuller DE(ed) Otolaryngology Head & Neck surgery. 3rd Ed St. Louis : Mosby Year Book : 19983294-3314.

[3]. Boneville F, Sarrazin JL, Marsot Dupuch KJ, Cordoliani YS, Doyon D, BonevellieJ F: Unusual lesions of the CPA: a segmental approach , Radiographics 2001;21: 419-43.

[4]. Verghese J, Mahore A, Goel A. Arachnoid cyst associated with painful tic convulsive. J. Clinical Neurosci; 19(2012)763-4.

[5]. Cirricillo SF, Cogen PH, Harsh GR, Edwards MS. Intracranial arachnoid cysts in children . A comparison of the effects of fenestration & shunting. J. Neurosurg;74(1991) 230-5.

[6]. Babu R, Murali R. Arachnoid cyst of CPA manifesting as contralateral trigeminal neuralgia: Case Report, Neurosurgery 1991; 28: 886-7.

[7]. Hayden MG, Tornabene SV, Nguyen A, Thekedi A, Alksene J F. Cerebellopontine angle cyst compressing the vagus nerve: Case Report. Neurosurgery 2007; 60: E1150.

[8]. Rasmussen P. Persistent mirror movements : A Clinical study of 17 children, adolescents and young adults. Dev Med Child Neurol. 1993: 699-707.

[9]. Gailea C, Popa T, Billot S, Meneret A, Depienne C, Roze E. Congenital mirror movements ; A clue to understanding bimanual motor control. J. Neurol. 2011; 258: 1911-19.