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Chiari Malformation Type 1 And Syringomyelia Together with a Colloid Cyst and Hydrocephalus

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Background: Chiari malformation (type 1) is a rare condition in which the cerebellar tonsils descend through the foramen magnum into the spinal canal. This in turn leads to compression of the spinal cord at foramen magnum interfering with the flow of cerebrospinal fluid leading to craniospinal dissociation and as a result hydrocephalus and syringomyelia. Sometimes it occurs secondary to space occupying lesions in intracranial compartments.

Key Words: Colloid Cyst, Chiari 1 Malformation, Hydrocephalus, Midline Suboccipital Craniotomy

Type 1 Chiari Malformation, also known as primary cerebellar ectopia or adult Chiari Malformation (CM) is a heterogeneous entity with the common feature of impaired cerebrospinal fluid circulation through the foramen magnum[1]. It is described as caudal displacement of cerebellum at least 5 cm below foramen magnum into the spinal canal without the involvement of brainstem[2] affecting roughly one in a thousand people symptomatically[3]. This pathology is most commonly associated with syringomyelia[4, 5]. Other conditions associated with CM include hydrocephalus, spina bifida, scoliosis[6] and basilar invagination[5].

A colloid cyst is a slow growing benign tumor which usually occurs in the anterior 3rd ventricle. They are known to be developed from primitive neuroepithelium or endoderm and contain a gelatinous material inside[7]. It represents less than 2% of all primary brain tumors [8]. Hydrocephalus is usually due to venous distensions and decreased absorption due to disturbances in CSF flow mechanics from cranial to spinal causing changes in pressure dynamics. About 6.5%-9.6% people with CM develop hydrocephalus[9]. We present a rare case of a patient with association of Colloid Cyst with Type 1 Chiari Malformation which presented a dilemma whether or not the hydrocephalus was Chiari associated or obstructive type from colloid cyst.

Case Description:

A 59-year-old female presented to the neurosurgery outpatient department with headache, neck and upper limb pain. Neurological examination revealed left sided cerebellar signs. Her deep tendon reflexes of the upper limb were slightly exaggerated (3+). Magnetic Resonance Imaging (MRI) of the brain and cervical spine showed tonsillar herniation into the upper cervical canal up to 11 mm suggestive of Chiari type 1 malformation and an associated colloid cyst measuring around

1cm with obstructive hydrocephalus. Shown in Figure1(a, b, c and d).

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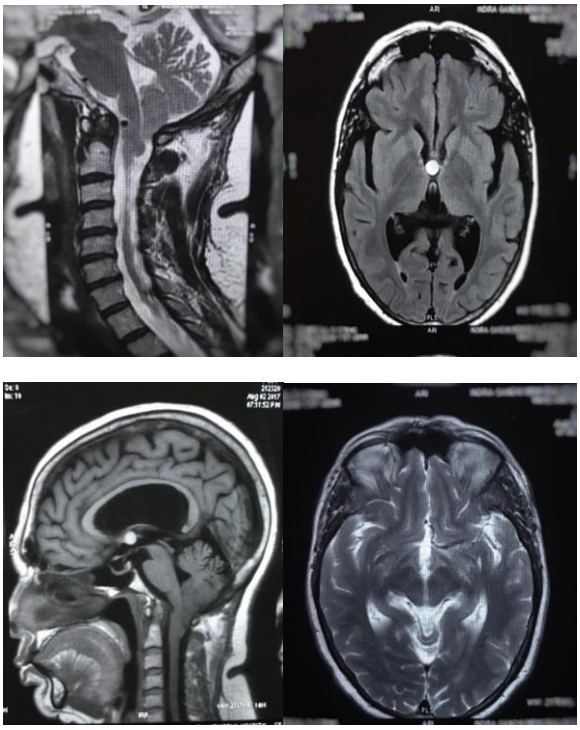

Figure 1(a): T2 weighted MRI Sagittal: tonsillar herniation and syringomyelia, (b): T1 weight MRI Axial: colloid cyst in the 3rd ventricle, (c): T1 weight MRI Sagittal: colloid cyst in the 3rd ventricle, (d): T2 weighted MRI Axial: Hydrocephalus. Visible temporal horns and sylvian fissure

Based on the clinical features and radiologic findings a diagnosis of Chiari Type 1 Malformation with colloid cyst causing hydrocephalus was made. She underwent endoscopic colloid cyst excision as the first procedure in August 2017. Post colloid cyst excision she had persistent symptoms of posterior fossa compressive signs. Hence, she underwent Midline sub occipital craniotomy (MSOC) and decompression of Foramen Magnum as her second procedure in December 2017.

Almost one month after MSOC (January 2018), she presented with headache, blurring of vision, right side swaying while walking and difficulty sleeping. On examination, left sided dysdiadochokinesia was present. MRI revealed right frontal tract gliosis, hydrocephalus, tonsillar descent and cervical syrinx. There was no evidence of colloid cyst or no periventricular ooze (Figure 3 and b).

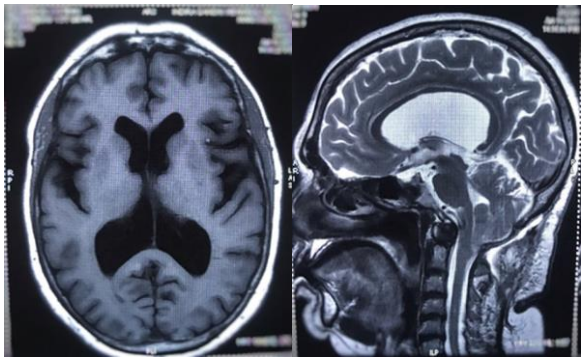

Figure 3 (a): T1 weighted Axial: colloid cyst not seen, (b): T2 weighted Sagittal: tonsillar descent and syrinx still present seen

In June 2019, she came to the outpatient department with complaints of memory loss, headache and dizziness which worsened while coughing and sneezing and additionally loss of balance. On examination she had left sided cerebellar signs and fundoscopy positive for papilledema. Lumbar puncture manometry showed a pressure of 34cm of cerebrospinal fluid. Therefore, CSF diversion with ventriculoperitoneal (VP) shunting was performed.

After the VP shunt, she followed up for regular checkup and her headache was improving. However, her cerebellar signs on the left side were positive and Hoffman's test was also positive. Follow-up MRI brain taken one year after (in 2020) showed the presence of syrinx but no signs of hydrocephalus or colloid cyst and there was no cerebellar pegging as shown in Figure 4 (a and b).

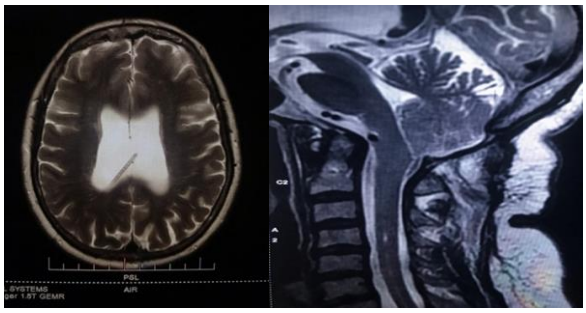

Figure 4 (a): T2 weighted Axial: VP shunt, (b): T2 weighted Sagittal: No cerebellar peg present

On her last follow up which was on 12th June 2022 she had complaints of only headache. Her cerebellar signs were negative and gait had improved. When compared with her initial clinical

presentation her signs and symptoms had improved significantly after the necessary interventions.

Discussion

Type 1 Chiari Malformation usually as a result of a smaller posterior fossa, mostly congenital[5]. In addition to this, they can also be due to intracranial lesions such as an arachnoid cyst, meningioma which can increase the intracranial pressure or can also occur following traumatic lumbar puncture and lumbo-peritoneal shunting[13]. If the clinical features in a Chiari type 1 malformation becomes apparent for the first time in old age, it may suggest the existence of an acquired intracranial mass[12]. Sakaki et al described 3 cases, one of which was a 65-year-old male who presented with gait disturbance and visual blurring. Further examination revealed nystagmus, dystaxia of all 4 extremities and mild right hemiparesis. Investigations showed meningioma in the right parietal region and cerebellar tonsillar herniation. The 2nd case was about a 55-year-old male who presented with neurological signs and symptoms and CT showed a subdural hematoma and tonsillar herniation. The last case was a 69-year-old male who presented with almost similar complaints was found to have right sided frontal meningioma with herniated cerebellar tonsils. All of these patients underwent surgery for the supratentorial lesion as well as decompression for Chiari malformation as excision of supratentorial lesion alone did not improve the symptoms completely. Following both procedures their clinical features are resolved[12]. Similarly, in our case there was a supratentorial mass, a colloid cyst, located in the anterior aspect of the 3rd ventricle. In addition to this, excision of colloid alone did not improve all the clinical features and she had to undergo MSOC in our case as well.

Due to the delicate location of origin of the colloid cyst, it often obstructs the foramina of Monro. Foramina of Monro connects the third ventricle with the lateral ventricle and acts as passageway for cerebrospinal fluid to flow out of the lateral ventricle into the third ventricle. Consequently, it may result in obstructive hydrocephalus and ventriculomegaly[8]. Therefore, it is possible that the tonsillar herniation in this patient is being caused by hydrocephalus linked to the colloid cyst.

Colloid cysts are typically found by chance and symptoms are frequently brought on by hydrocephalus[8]. Headache, nausea, vomiting, lethargy, gait disturbances and blurred vision are some of the symptoms[14]. In this patient also there were many features suggesting non-communicating hydrocephalus such as headache, gait disturbances, blurring of vision. Taking all these into account, Chiari type 1 malformation could have been a congenital cause or acquired both in this case as well as in the cases described by Sakaki et al[12].

Colloid cyst has been described as a sudden cause of death due to acute obstruction of CSF flow[15]. Few cases of sudden death associated with colloid cyst have been reported by15, 16, 17. Therefore it is vital to eliminate this risk by firstly operating on the colloid cyst.

Syringomyelia is another condition that was found in this patient. Syringomyelia is caused due to the presence of a fluid filled cavity within the spinal cord[18]. Syringomyelia can occur due to various reasons such as following an infection of the spinal cord, post trauma[19]. However, most of the time it is associated with Chiari 1 malformation[5]. Syringomyelia in Chiari malformations are usually due to disturbances in cerebrospinal fluid (CSF) flow between cranium and spinal column usually during cardiac systole forcing CSF into the central canal with each arterial pulse resulting in dilation of central canal (Gardener's water hammer theory)[20]. Majority of the time Syringomyelia can cause pain, progressive weakness and muscle wasting and hypoesthesia in the upper limb. It can also cause headaches and stiffness in lower limbs.

It is challenging to determine if the headaches, neck and upper limb pain and all the neurological deficits in this patient were majorly caused by the colloid cyst or the congenital tonsillar herniation or combination of the two.

The mechanism of tonsillar herniation in our case could be similar to that explained by most authors, a result of interaction between mechanical and embryonic factors. The effect of a mass occupying lesion in the infratentorial or supratentorial spaces may promote the development of congenital cerebellar tonsil herniation[12].

Confirming whether the Chiari malformation in this patient is acquired or congenital and the treatment prioritization is one of the most challenging decisions for the surgeon. One can argue that the tonsillar herniation was caused due to intracranial hypertension caused by the colloid cyst and tonsillar herniation could be the cause for syringomyelia[21]. However, it is rare for Chiari malformation and syringomyelia to be caused by a colloid cyst with hydrocephalus[10].

Furthermore, removal of the intracranial tumor did not cause resolution of the hydrocephalus, tonsillar herniation and syringomyelia. Therefore, altered CSF dynamics and resorption due to obstruction by colloid cyst was not the only factor contributing to the development of these pathologies.

A case report written by Thotakura et al[10] describes a similar case about a patient who had Chiari 1 malformation with syringomyelia with colloid cyst and hydrocephalus. In this case the tonsillar herniation and the syringomyelia got resolved after the excision of the colloid cyst making the diagnosis more likely Acquired chiari 1 malformation with syringomyelia secondary to colloid cyst with hydrocephalus. Therefore, in this case MSOC was not required. Another study showed that the percentage of tonsillar ascent, syrinx resolution and neurologic recovery were equal in patients who underwent lesion excision alone and those who underwent posterior fossa decompression and lesions removal[11]. Sakaki et al described 3 cases which had a supratentorial mass as well as cerebellar herniation. All these 3 cases required surgery for the lesion and decompression on the Chiari malformation for the complete resolution of symptoms.

The persistence of tonsillar herniation, cerebellar signs and non-resolution of hydrocephalus and syringomyelia could be due to inadequate CSF follow dynamic despite sufficient decompression of Foramen Magnum owing to postoperative adhesion as a factor. A study published by Shweikeh et al found that hydrocephalus was one of the most common complications following posterior fossa decompression[22]. Study conducted by Jian et al found that younger patients, patients with excessive blood loss intraoperatively and those having a fourth ventricular web were more likely to develop hydrocephalus following posterior fossa decompression[23].

Additionally, surgeons need to prioritize the order of treatment accordingly to prevent life threatening complications such as sudden death recorded in colloid cysts of the third ventricle. It is noteworthy that the initial surgical treatments including colloid cyst excision, posterior fossa decompression improved symptoms to a certain extent, but did not resolve the most pressing issue of papilledema. Hence, she was planned to undergo lumbar puncture manometry. After lumbar puncture her condition improved but she was still having on and off pain and an MRI brain showed hydrocephalus. Finally, she underwent right occipital ventriculoperitoneal (VP) shunting. Following VP shunting however had a significant impact on lowering the intracranial pressure and in turn improving papilledema and her symptoms resolved to near normal within 1-2 years.

Conclusion:

Chiari type 1 malformation with colloid cyst causing obstructive hydrocephalus is a rare presentation but rather substantial pathology to be identified by the neurosurgeons. At times it may be difficult to identify the primary cause of hydrocephalus and tonsillar herniation. One can argue that it is the colloid cyst causing the obstruction of the CSF pathway and thus leading to tonsillar herniation. On the contrary, Chiari itself may be the cause of hydrocephalus. In this case it is more reasonable to say that colloid cyst was an accidental association and has less contribution to the signs and symptoms. Theoretically though colloid was excised with establishment of clear CSF ventricular pathway, surgery itself may have led to intraventricular perioperative bleed related communicating hydrocephalus which did not allow the intracranial pressure to resolve despite flow creation. Therefore, in such complex cases it is important for the neurosurgeons to be vigilant in order to follow up these patients closely to not miss a fatal change in course of illness which may lead to sudden deterioration of the patient's condition. Though this particular case didn't attain the expected result following the first two surgeries it is important to stress that prioritization of one procedure over the other would be essential in

order to avoid possible major complications related to certain pathologies.

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