Large tentorium meningioma causing chiari malformation type-1 with syringomalia with complete resolution of syrinx and chiari after surgical excision: rare case report with review of literature

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Abstract: A 35-year-old female was admitted with a 3-year history of headache, gait disturbance and vertigo on & off and one year history of nasal regurgitation. Magnetic resonance imaging demonstrated a large tentorium meningioma left sided and syringomyelia in the upper cervical cord associated with caudal displacement of the cerebellar tonsil (chiari type -1 malformation). Herniation of the cerebellar tonsil and distortion of the brain stem had probably caused disturbance of cerebrospinal fluid flow which combined with obstruction of the spinal canal, caused the syrinx. Complete excision of the tumor resulted in symptomatic improvement of these symptoms with complete resolution of syrinx & chiari.

Key words: syringomyelia, posterior fossa, brain tumor, meningioma, Chiari malformation

Introduction

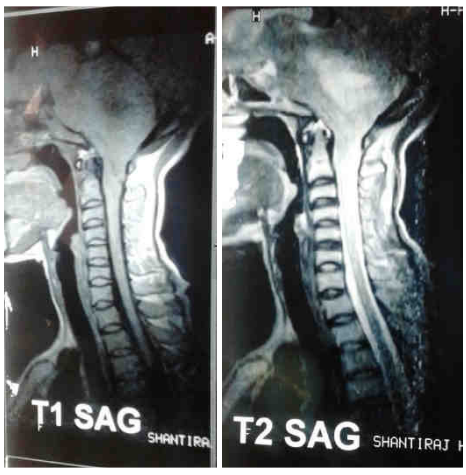
Chiari & Syringomyelia is well-known to be associated with anomalies in the craniocervical junction. The combination of Chiari type-1 with syringomyelia and posterior fossa tumor is rare. We Describe a case of syringomyelia with chiari malformation (CM) type -1 associated with Tentorial meningioma and discuss the pathogenesis based on neuroimaging findings.

Case report

A 36-year-old female was admitted with complaints of suboccipital headache, vertigo on & off and gait difficulty persisting for the last 3 years and nasal regurgitation for last 1 year. Neurological examination revealed left cerebellar sign present with nystagmoid ocular movement and hypertonia in both lower limb and power all four limbs 5/5 except both hand grip 80%. All deep tendon reflexes exaggerated with bilateral knee clonus present. Magnetic resonance (MR) imaging with gadolinium-diethylenetriaminepenta-acetic acid also demonstrated a homogeneously enhanced extra axial lesion in the Left

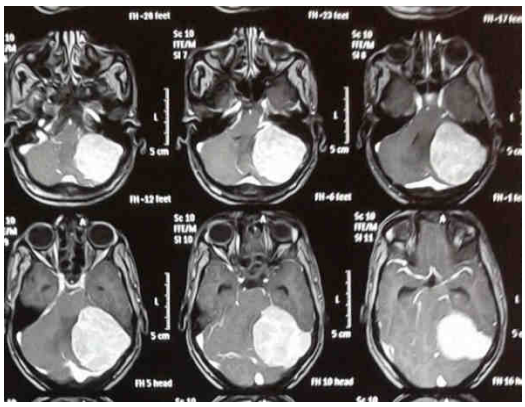
tentorium causing mass effect on left cerebellar hemisphere with effacement of fourth ventricle resulting dilatation of supra tentorial ventricle with tonsilar herniation. Sagittal MR imaging indicated a peg-like deformity of the cerebellar tonsil and descend up to C2 with syringomyelia of the upper cervical region of spinal cord (Figures 1, 2). Midline suboccipital craniectomy with extending left suboccipital craniectomy with foramen magnum decompression with Simpson grade II excision of meningioma was done. Histological examination showed Transitional meningioma.

The postoperative course was uneventful. MR imaging done 6 months after the surgery does not show any caudal displacement of tonsil and complete resolution of syringomyelia (Figure 3). Patient is completely asymptomatic at present.



A B

Figure 1 A&B - T1 & T2 weighted sagittal MR image showed a descent and plugging of the cerebellar tonsils up to C2 with presence of a syringomyelia extending from the cervicomedullary junction till C4 segment. However, some signal changes probably suggestive of myelomalacia



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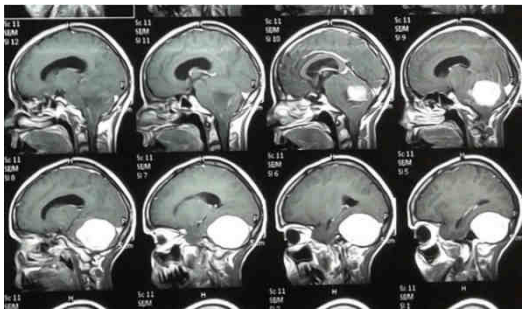
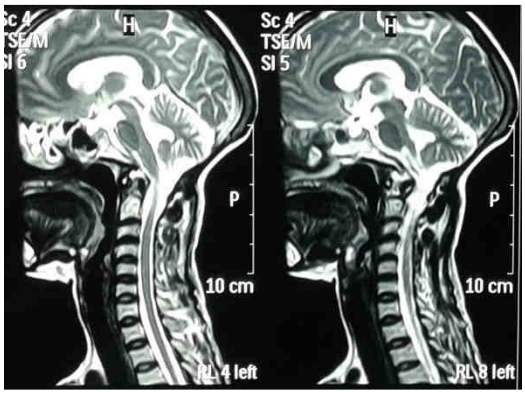


B

Figure 2A - T1 weighted axial MRI brain with contrast revealed extra axial mass lesion in Left cerebellar hemisphere along tentorium with effacement of forth ventricle

Figure 2B - T1 weighted sagittal MRI brain with contrast revealed extra axial mass lesion along left tentorium with cerebellar tonsil herniation up to C2



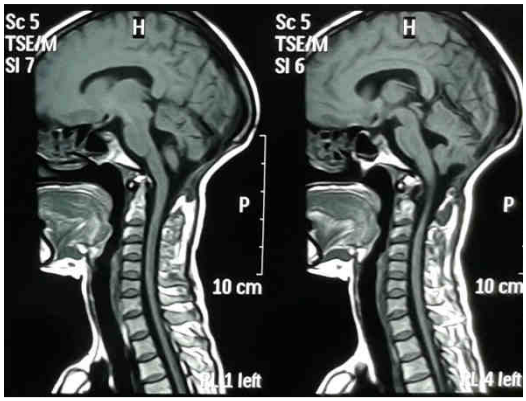


Figure 3 - T2 & T1 weighted sagittal MRI brain with spine revealed complete resolution of syrinx and tonsilar herniation

Discussion

Posterior fossa tumors associated with syringomyelia include brainstem glioma, (20) meningioma, (9, 12, and 16, 10, 7, 2) Cerebellar astrocytoma, (13) cerebellar hemangioblastoma, (6) and fourth ventricular epidermoid tumor (11).

There are several theories to explain the pathophysiology of syringomyelia. Gardner's (8) suggested that a congenital imperforate of Magendie's foramen disturbs the cerebrospinal fluid flow out of the cisterna magna and an intracranial arterial pulse produces a water-hammer effect on the central canal, leading to enlargement of the syrinx. Williams and Timperley (20) stressed the importance of craniospinal pressure dissociation, with the venous pressure change occurring soon after coughing evacuating the central canal with syrinx extension. The check valve effect associated with foramen magnum obstructive lesions may enhance syringomyelia, (19) a theory accepted by many clinicians. Ball and Dayan (4) found that cerebrospinal fluid leaking into the spinal cord along Virchow-Robin spaces may cause syringomyelia. Aubin et al. (3) observed the transneural passage of cerebrospinal fluid into the spinal cord by comparison of the CT density of the subarachnoid space, spinal cord, and syringomyelic cavity. Barnett et al. (5) classed syringomyelia into communicating and non-communicating types. Communicating syringomyelia is consistent with Gardner's hydrodynamic theory, with communication between the syrinx and fourth ventricle. Non-communicating syringomyelia is secondary to intramedullary tumors or spinal injury, with no communication between the syrinx and fourth ventricle. Various pathogeneses for syringomyelia with posterior fossa tumor have been identified. De Reucket al. (6) reported syringomyelia with cerebellar hemangioblastoma and concluded that faulty closure of the dorsal raphe with glial inclusion caused the syrinx. Williams and Timperley (20) reported three cases of syringomyelia with brainstem glioma and emphasized the importance of craniospinal pressure dissociation and evacuation of the central canal.

Neuroimaging of the present case revealed caudal displacement of the lower cerebellar tonsil with the same configuration as that of Chiari type I malformation. However, removal of the tumor resulting the cerebellar tonsil to return to its normal position. This observation suggests that the cause of syringomyelia must be an anatomical change around the craniocervical junction. Several theories have been proposed to explain the development of syringomyelia. Failure of the foramina of the

fourth ventricle to open with continuing communication between the fourth ventricle and the cystic space within the spinal cord via the obex may allow increased pressure within the ventricles to be transmitted to the central canal (8). CSF pressure waves can cause forced flow of the CSF into the syrinx along the Virchow-Robin spaces (4). Partial blockage of the subarachnoid space in the region of the cisterna magna may direct CSF into the communication, providing an intermittent distending force which may be active for many years (18). Obstruction at the cisterna magna associated with a high venous pressure can lead to transmedullary passage of CSF which could create a syrinx cavity (1). Excessive absorption of CSF from the spinal cord might cause Chiari type I malformation, leading to a foraminal obstruction and ultimately producing a syrinx (17). Recently, a detailed analysis of the configuration of the central canal in the normal population as well as in syringomyelia patients showed that a large portion of the normal group has an obstructed central canal except in their early stages of life. The spinal cord with syrinx shows three patterns of communication with the fourth ventricle and central canal: central canal syrinx (communicating), central canal syrinx (non-communicating), and extracanalicular syrinx. Central canal syrinx (communicating) is observed predominantly in children with hydrocephalus. Central canal syrinx (non-communicating) has a cavity consisting of a focal dilation of central canal that is separated from the fourth ventricle by a syrinx-free segment of spinal cord and occurs predominantly in adult patients with various diseases which cause CSF circulatory disturbance around the cervicomedullary junction. Extracanalicular syrinx is seen as a result of spinal cord injury. MR images of patients with brain tumors which demonstrate syrinx may suggest that the central canal has already been occluded in some locations. Phase-contrast/cine MR imaging indicates that a disturbance of CSF circulation in the spinal subarachnoid space may cause fluid to be forced into the central canal through the interstitial space of the spinal cord in such cases (14). Syringomyelia may occur secondary to the brain tumor within the posterior fossa but without symptoms suggestive of syringomyelia. (2, 7, 9, 10, 12, 13, 15, 16, 20) Sagittal MR imaging of our case showed a large and slow-growing brain tumor, which had resulted in elimination of CSF from the posterior fossa and herniation of the cerebellar tonsil through the foramen magnum. Herniation of the cerebellar tonsil and distortion of the lower brain stem may have disturbed CSF circulation in the spinal subarachnoid space, and resulted in transmedullary passage of CSF. Furthermore, the obstructed central canal may also have prevented free passage of CSF outside the central canal. Consequently, these pathological CSF flows and accumulations caused a compartment of the central canal force to dilate. The symptoms of brain tumor in the posterior fossa, as with all cases with tonsilar herniation, are too severe to remain untreated. Syringomyelia with CM type-1 associated with various intracranial diseases can be diagnosed by neurological imaging. An early diagnosis is essential for a good prognosis.

Conclusion

Extra-axial tumor of posterior fossa is rarely associated with syringomalia & Chiari-malformation. Surgical excision of the primary lesion (Tumor) resulting complete resolution of syrinx & Chiari.

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