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Research Article

CHIARI MALFORMATION TYPE-1 AND SYRINGOMYELIA ASSOCIATED WITH LEFT CEREBELLAR EPIDERMOID TUMOR: A RARE CASE REPORT

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A B S T R A C T

Chiari malformation (CM) is a structural defect in the cerebellum, characterized by a downward displacement of one or both cerebellar tonsils through the foramen magnum and Syringomyelia is a chronic progressive degenerative disorder characterized by a fluid-filled cyst located in the spinal cord. Syringomyelia is often associated with type I Chiari malformation and is commonly seen between the C-4 and C-6 levels. The exact development of syringomyelia is unknown but many theories suggest that the herniated tonsils in type I Chiari malformations cause a "plug" to form, which does not allow an outlet of CSF from the brain to the spinal canal. Syringomyelia is present in 25% of patients with type I Chiari malformations. On the other side, intracranial epidermoid tumor is relatively common congenital lesion wherean inclusion of displaced dorsal midline ectodermal cell rests occurs during neural tube closure. We found a 14-year-old boy with spastic quadriparesis with difficulty in walking with poor balance for 6 months. Magnetic resonance imaging demonstrated a large epidermoid in left cerebellum and syringomyelia extending from C4 to D4 associated with caudal displacement of the cerebellar tonsil (chiari type -1 malformation). A Midline suboccipital craniectomy with extending left suboccipital craniectomy with foramen magnum decompression along with excision of posteior arch of C1 and complete removal of Epidermoid Tumour with expansile duroplasty was done.

After operation, resulted in symptomatic improvement of these symptoms with complete resolution of syrinx & chiari. After surveying the literature, we think, we have found the rarest association, an epidermoid tumor with chiari malformation type I & syringomyelia.

INTRODUCTION

Chiari malformation & 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 epidermoid tumor and the pathogenesis based on neuroimaging findings.

Chiari malformation (CM) is a structural defect in the cerebellum, characterized by a downward displacement of one or both cerebellar tonsils through the foramen magnum (the opening at the base of the skull).The malformation is named for Austrian pathologist Hans Chiari. In the late 19th century, Austrian pathologist Hans Chiari described seemingly related anomalies of the hindbrain, the so-called Chiari malformations I, II, III, IV. The scale of severity is rated I – IV, with IV being the most severe and not compatible with life. Types III and IV are very rare.[1,2,11,13]

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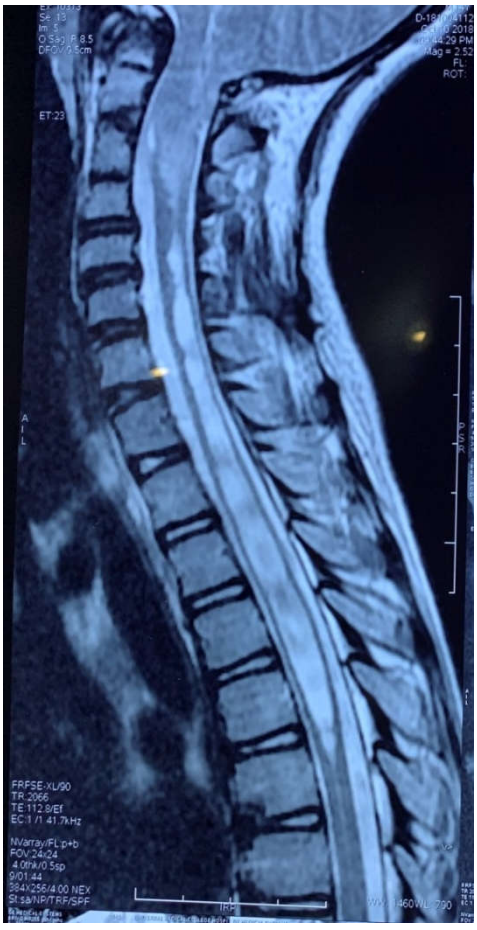
Syringomyelia is a chronic progressive degenerative disorder characterized by a fluid-filled cyst located in the spinal cord. Its symptoms include pain, weakness, numbness, and stiffness in the back, shoulders, arms or legs. Other symptoms include headaches, the inability to feel changes in the temperature, sweating, sexual dysfunction, and loss of bowel and bladder control. It is usually seen in the cervical region but can extend into the medulla oblongata and pons or it can reach downward into the thoracic or lumbar segments. Syringomyelia is often associated with type I Chiari malformation and is commonly seen between the C-4 and C-6 levels. The exact development of syringomyelia is unknown but many theories suggest that the herniated tonsils in type I Chiari malformations cause a "plug" to form, which does not allow an outlet of CSF from the brain to the spinal canal. Syringomyelia is present in 25% of patients with type I Chiari malformations.[3,4]

On the other side, Intracranial epidermoid cysts are relatively common congenital lesions which account for about 1% of all intracranial tumors. It is an inclusion of displaced dorsal midline ectodermal cell rests during neural tube closure.

Though syringomyelia is commonly associated with Chiari malformation , other conditions sometimes associated with Chiari malformation include hydrocephalus, spinal curvature, tethered spinal cord syndrome, and connective tissue disorders such as Ehlers-Danlos syndrome and Marfan syndrome andplatybasia (flattening of the skull base).[13,15] So, in this case, we think, we have found the rarest association, an epidermoid cyst.

Case Study

A 14-year-old boy was admitted with spastic quadriparesis with difficulty in walking along with poor balance and occipital headache for 6 months. On neurological assessment, we found loss of pain and temperature sensation in upper limbs with upper trunk, 3/5 grade power (MRC grading) in upper limb and 4/5 grade power (MRC grading) in lower limb muscles bilaterally and exaggerated deep tendon reflexes. Magnetic resonance imaging demonstrated a large epidermoid in left cerebellum and syringomyelia extending from C4 to D4 associated with caudal displacement of the cerebellar tonsil (chiari type -1 malformation). A Midline suboccipital craniectomy with extending left suboccipital craniectomy with foramen magnum decompression along with excision of posteior arch of C1 and complete removal of Epidermoid Tumour with expansile duroplasty was done. Histopathological analysis confirmed an epidermoid tumour. After operation, resulted in symptomatic improvement of these symptoms with complete resolution of syrinx & chiari.



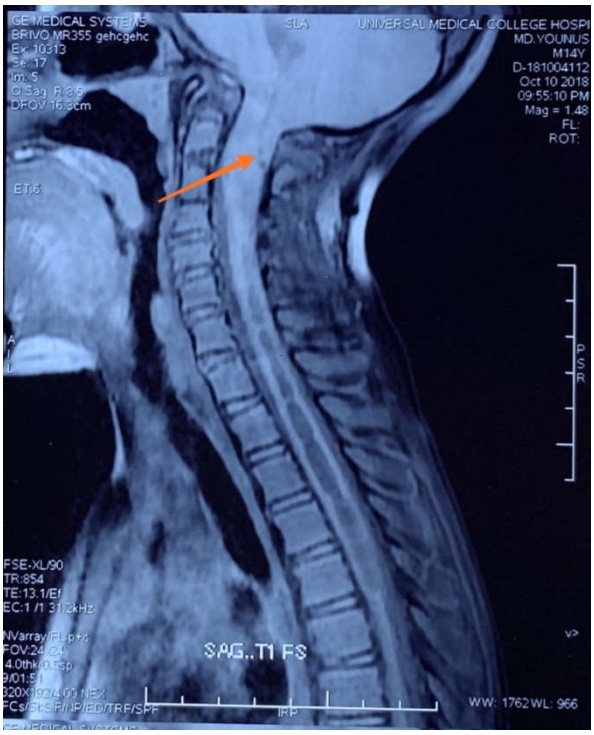


Fig 1 A & B T2 & T1 weighted sagittal MRI showed decent and plugging cerebellar tonsil upto C2 with syringomyelia extending to C4 to D4

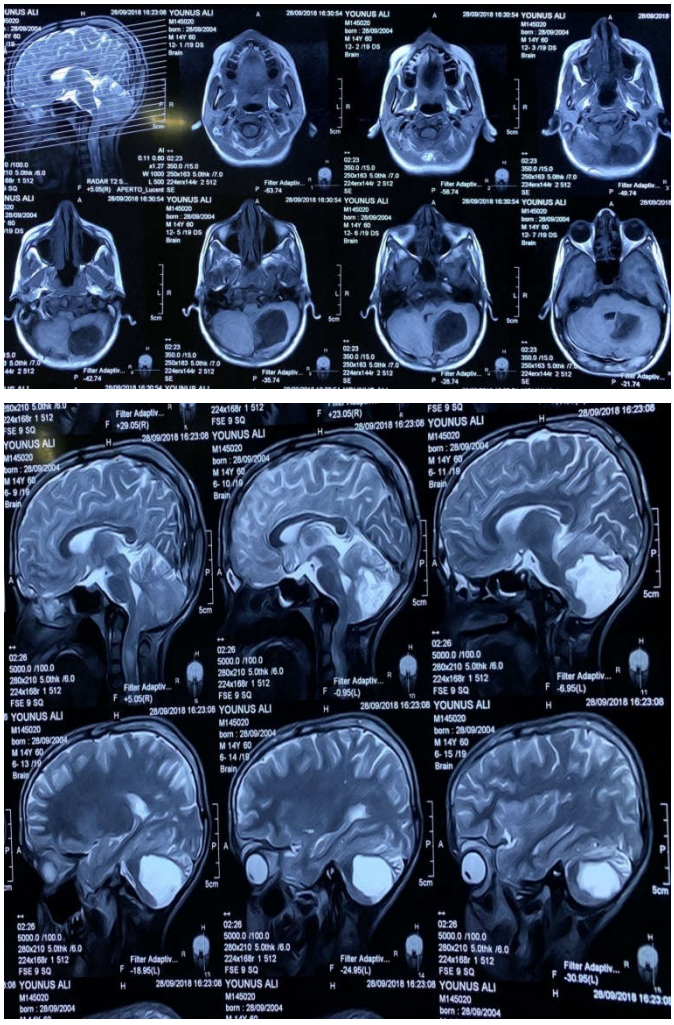


Fig 2 A & B: T1 & T2 weighted MRI of brain showed left cerebellar epidermoid

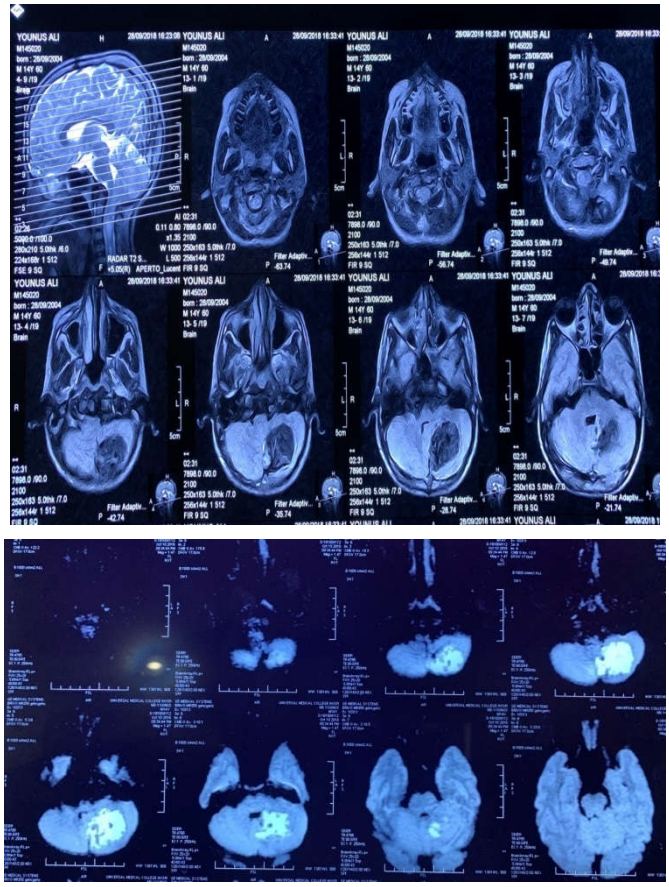


Fig 3 A & B: T1 weighted contrast axial MRI & DWI showed left cerebellar epidermoid

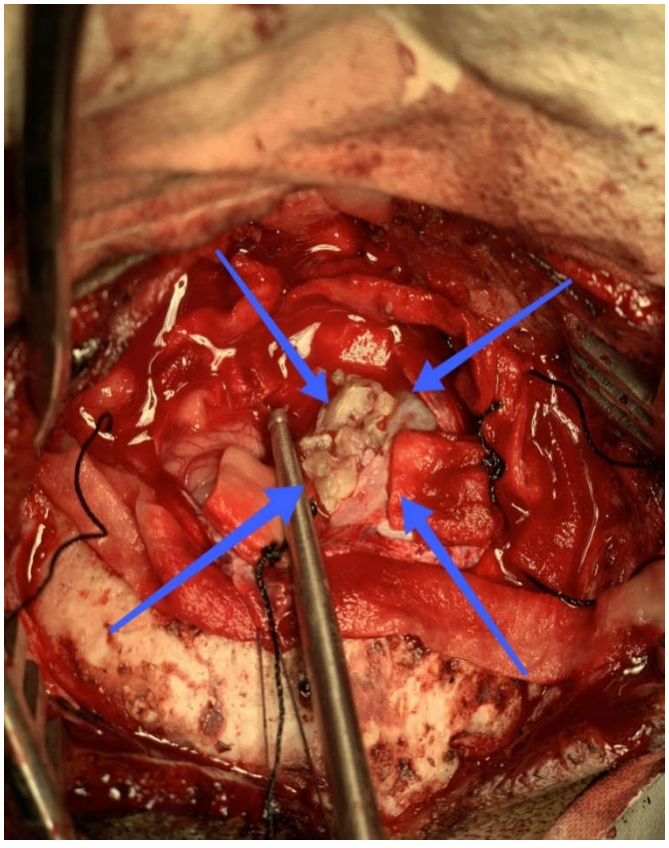
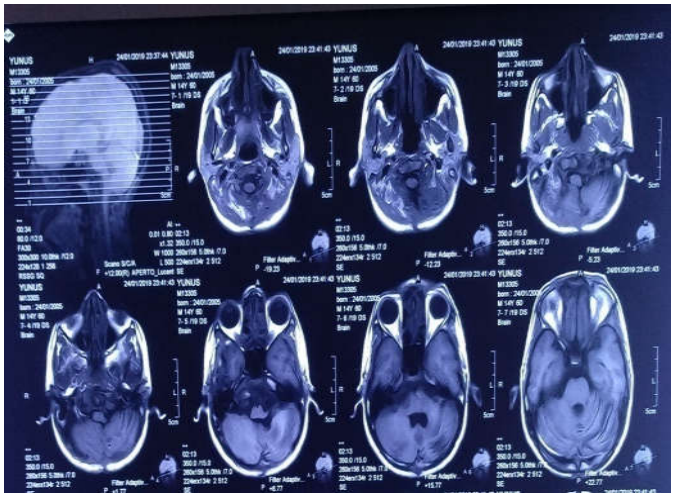


Fig 4 Peroperative view of Epidermoid tumour



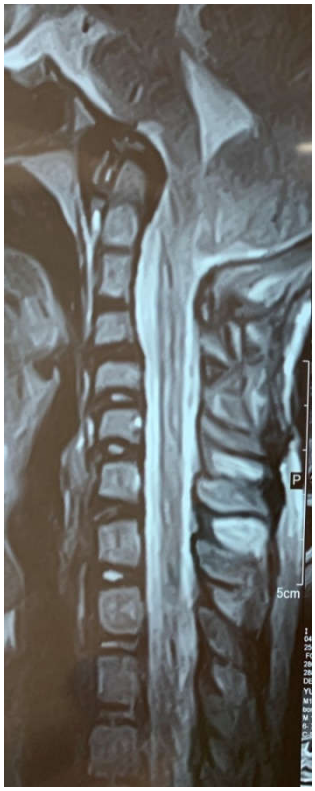


Fig 5 A & B Postoperative T1 weighted contrast axial MRI of brain and T2 sagittal MRI showed complete removal of epidermoid with resolution of syrix and tonsillar herniation

DISCUSSION

Chiari malformation (CM) is a structural defect in the cerebellum, characterized by a downward displacement of one or both cerebellar tonsils through the foramen magnum (the opening at the base of the skull). The malformation is named for Austrian pathologist Hans Chiari. In the late 19th century, Austrian pathologist Hans Chiari described seemingly related anomalies of the hindbrain, the so-called Chiari malformations I, II, III, IV. The scale of severity is rated I – IV, with IV being the most severe and not compatible with life. Types III and IV are very rare.[1,11,13]

Congenital Chiari I malformation, defined as tonsilar herniations of 3 to 5 mm or greater, was previously believed to be in the range of one per 1000 births, but is likely much higher. Women are three times more likely than men to have a congenital Chiari malformation. A study using upright MRI found cerebellar tonsillar ectopia in 23% of adults with

headache from motor-vehicle-accident head trauma. Upright MRI was more than twice as sensitive as standard MRI, likely because gravity affects cerebellar position.[11,12,13]

The most widely accepted pathophysiological mechanism by which Chiari type I malformations occur is by a reduction or lack of development of the posterior fossa as a result of congenital or acquired disorders. Congenital causes include hydrocephalus, craniosynostosis (especially of the lambdoid suture), hyperostosis (such as craniometaphyseal dysplasia, osteopetrosis, erythroid hyperplasia), X-linked vitamin D-resistant rickets, and neurofibromatosis type I. Acquired disorders include space occupying lesions due to one of several potential causes ranging from brain tumors to hematomas. Head trauma may cause cerebellar tonsillarectopia, possibly because of duralstrain. Additionally, ectopia may be present but asymptomatic until whiplash causes it to become symptomatic.[15,18]

Syringomyelia is a chronic progressive degenerative disorder characterized by a fluid-filled cyst located in the spinal cord. Its symptoms include pain, weakness, numbness, and stiffness in the back, shoulders, arms or legs. Other symptoms include headaches, the inability to feel changes in the temperature, sweating, sexual dysfunction, and loss of bowel and bladder control. It is usually seen in the cervical region but can extend into the medulla oblongata and pons or it can reach downward into the thoracic or lumbar segments. Syringomyelia is often associated with type I Chiari malformation and is commonly seen between the C-4 and C-6 levels. The exact development of syringomyelia is unknown but many theories suggest that the herniated tonsils in type I Chiari malformations cause a "plug" to form, which does not allow an outlet of CSF from the brain to the spinal canal. Syringomyelia is present in 25% of patients with type I Chiari malformations.[3,4]

Till now, there are a lot of debates about the pathoembryology of Chiari malformation type I. Cases of congenital Chiari malformation may be explained by evolutionary and genetic factors. Typically, an infant's brain weighs around 400g at birth and triples to 1100-1400g by age 11. At the same time the cranium triples in volume from 500 cm3 to 1500 cm3 to accommodate the growing brain. During human evolution, the skull underwent numerous changes to accommodate the growing brain. The evolutionary changes included increased size and shape of the skull, decreased basal angle and basicranial length. These modifications resulted in significant reduction of the size of the posterior fossa in modern humans. In normal adults, the posterior fossa comprises 27% of the total intracranial space, while in adults with Chiari Type I, it is only 21%. If a modern brain is paired with a less modern skull, the posterior fossa may be too small, so that the only place where the cerebellum can expand is the foramen magnum, leading to development of Chiari Type I.[11,18]

Type I, Herniation of cerebellar tonsils. Tonsillarectopia below the foramen magnum, with greater than 5 mm below as the most commonly cited cutoff value for abnormal position (although this is considered somewhat controversial). Syringomyelia of cervical or cervicothoracic spinal cord can be seen. Sometimes the medullary kink and brainstem elongation can be seen. Can be congenital, or acquired through trauma. When congenital, may be asymptomatic during childhood, but often manifests with headaches and cerebellar symptoms. Syndrome of occipitoatlantoaxial hypermobility is an acquired Chiari I malformation in patients with hereditary disorders of connective tissue. Patients who exhibit extreme joint hypermobility and connective tissue weakness as a result of Ehlers-Danlos syndrome or Marfan syndrome are susceptible to instabilities of the craniocervical junction; thus they are at risk for acquiring a Chiari malformation.

Type II, This is the only type also known as an Arnold-Chiari malformation. As opposed to the less pronounced tonsillar herniation seen with Chiari I, there is a larger cerebellar vermian displacement. Low lying torcularherophili (confluence of sinuses), tectalbeaking, and hydrocephalus with consequent clival hypoplasia are classic anatomic associations. Usually accompanied by a lumbar or lumbosacral myelomeningocelewithtonsillar herniation below theforamen magnum. The position of the torcularherophili is important for distinction from Dandy–Walker syndrome in which it is classically upturned. This is important because the hypoplastic cerebellum of Dandy–Walker may be difficult to distinguish from a Chiari malformation that has herniated or is ectopic on imaging. Colpocephaly may be seen due to the associated neural tube defect.

Type III, Associated with an occipital encephalocele containing a variety of abnormal neuroectodermal tissues. Syringomyelia and tethered cord as well as hydrocephalus is also seen.

Type IV, Characterized by a lack of cerebellar development in which the cerebellum and brain stem lie within the posterior fossa with no relation to the foramen magnum. Associated with hypoplasia and equivalent to primary cerebellar agenesis.[2,11,14,16,17]

In posterior fossa, epidermoid cysts grow through slow accumulation of desquamated epithelium between cranial nerves, vessels and the brain stem. [8,9,10] According to Gardner[3], inadequate permeability of the fourth ventricle outflow associated with blockage of cisterna magna and an unobstructed obex generate an open connection between the ventricular system and the spinal central canal, that ultimately causes development of a syrinx. In other theories, these lesions displace the posterior fossa structures caudally and create craniospinal and intraspinal pressure dissociation that leads to obstruction of the normal circulation of the CSF at the foramen magnum. Neurological dysfunction occurs when the obstruction persists as the accumulated fluid inside the spinal cord form a cavitation, causing expansion of the syrinx in craniocaudal and lateral direction. Therefore, our goal should be refurbishment of normal CSF circulation surgically that can ensured by posterior fossa and cervical decompression essentially. [4,5,6] Fuyou G *et al*. also[14] recommended for excision and removal of posterior fossa lesion and then observation of the influence of that procedure. Cervical syringomyelia also has been reported to be associated with meningioma, medulloblastoma, glioma, arachnoid cyst in the posterior fossa. [15] The same rationale should be applicable in those tumours. Though syringomyelia is commonly associated with Chiarimalformation, other conditions sometimes associated with Chiari malformation include hydrocephalus, spinal curvature, tethered spinal cord syndrome, and connective tissue disorders such as Ehlers-Danlos syndrome and Marfan syndrome and platybasia (flattening of the skull base).[5,6,7,15] After surveying the literature, we found one article 'Dermoid of the Posterior Fossa in Chiari II

Malformation: The First Reported Case'. In our case, after two months of posterior fossa and upper cervical decompression, the patient came to us with satisfaction about the improvement of his limb spasticity and weakness and resolution of headache. A follow up MRI of spine showed collapsed syrinx and totally absence of posterior fossa lesion. [19]

During the 10-year period from 1994 until May 2004, among the 435 patients with developmental CVJ four of these patients harbored a constellation of CVJ anomalies with dermoid and epidermoid cysts (hospital prevalence, 0.9%). All patients (ages 18, 23, and 25 yr) presented with features of spastic quadriparesis, restriction of neck movements, and raised intracranial pressure. Magnetic resonance imaging showed features of CVJ anomalies in all patients (occipitalization of C1, 3 patients; basilar invagination, 3 patients; atlantoaxial dislocation, 4 patients; and an abnormal posteriorly pointed dens, 1 patient), along with a Klippel-Feil anomaly (Patients 1–3**,** 2nd and 3rd cervical vertebrae). Patient 4 also had a Chiari malformation with syrinx. In addition, all four patients had coexisting dermoid or epidermoid cysts (Patients 1 and 3, midline posterior fossa epidermoid; Patient 2, midline posterior fossa dermoid; Patient 4, quadrigeminal cistern epidermoid).[20]

CONCLUSION

After discussion and reviewing the literatures and considering the underlying pathogenesis of this kind of rare association of an epidermoid tumour with a Chiari Malformation type I, this is obvious that the extirpation of the posterior fossa lesion, whatever it may be, alone can lead us to the desired goal of curing the patient and preventing the complications of the syrinx. After surveying the literature, we think, we have found the rarest association, an epidermoid tumour with chiari malformation & syringomyelia.

Conflicts of interest- There are no conflicts of interest.

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