Teaching Neuro*Images*: Acquired Chiari malformation with syringohydromyelia caused by posterior fossa tumor

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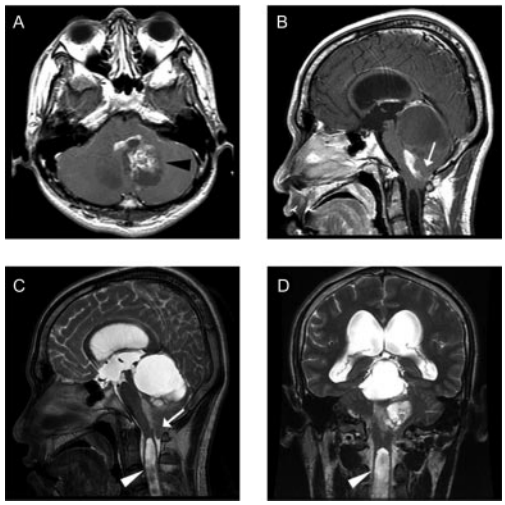
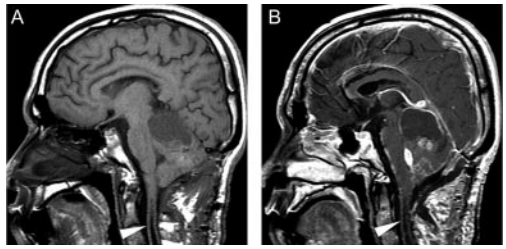
Figure 1 Acquired Chiari malformation with syringohydromyelia caused by cerebellar pilocytic astrocytoma



Gadolinium-enhanced axial (A) and sagittal (B) T1-weighted brain MRI showing a heterogeneous enhancing cystic tumor of posterior fossa (black arrowhead) compressing the fourth ventricle with marked hydrocephalus and tonsillar herniation (white arrow) down to the C2 level. Sagittal (C) and coronal (D) T2-weighted MRI showing severe cervical syringohydromyelia (white arrowhead) was centrally located from C1 to C3. Acquired Chiari malformation with cervical syringohydromyelia was diagnosed.

A 28-year-old man was admitted due to progressive headache and blurred vision. Neurologic examination showed bilateral papilledema. Brain MRI showed a cystic posterior fossa tumor compressing the fourth ventricle with tonsillar herniation and cervical syringohydromyelia (figure 1). The tumor was removed through a suboccipital craniectomy and pilocytic astrocytoma was diagnosed histologically. One month later, his symptoms had improved with shrinkage of syringohydromyelia by postoperative MRI (figure 2).

Figure 2 Follow-up postoperative brain MRI



Sagittal T1-weighted (A) and gadolinium-enhanced (B) brain MRI showing residual posterior fossa pilocytic astrocytoma with shrinkage of cervical syringohydromyelia (white arrowhead).

Acquired Chiari malformation with syringohydromyelia is a rare disease with the incidence of 3% in posterior fossa tumors.[1] The pathogenesis is thought due to blockage of CSF flow at the foramen magnum, causing disruption of the blood–brain barrier with syringohydromyelia formation most often at the cervical spine level. Acquired Chiari malformation with syringohydromyelia caused by posterior fossa tumors is usually asymptomatic.[1] Early diagnosis by brain MRI and prompt decompressive suboccipital craniectomy may prevent disease progression and improve prognosis.[2]

REFERENCES

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