Teaching NeuroImages: Acquired Chiari malformation with syringohydromyelia caused by posterior fossa tumor

Fu Z. Wu, MD
Jui H. Fu, MD
Jun Y. Chen, MD
Ping H. Lai, MD

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 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).

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


From the Departments of Radiology (F.Z.W., J.H.F., P.H.L.) and Neurosurgery (J.Y.C.), Kaohsiung Veterans General Hospital, Kaohsiung; and School of Medicine (F.Z.W., J.H.F., J.Y.C., P.H.L.), National Yang-Ming University, Taipei, Taiwan.

Study funding: Supported by the National Science Council (NSC-97-2314-B-075B-010-MY3) and Veterans General Hospital-Kaohsiung (VGHKS99-66).

Disclosure: Dr. Wu and Dr. Fu report no disclosures. Dr. Chen receives research support from the Veterans General Hospital-Kaohsiung. Dr. Lai receives research support from the National Science Council and the Veterans General Hospital-Kaohsiung.
Teaching NeuroImages: Acquired Chiari malformation with syringohydromyelia caused by posterior fossa tumor
Neurology 2010;75:e59
DOI 10.1212/WNL.0b013e3181f612fa

This information is current as of October 4, 2010

Updated Information & Services
including high resolution figures, can be found at:
http://www.neurology.org/content/75/14/e59.full.html

References
This article cites 2 articles, 0 of which you can access for free at:
http://www.neurology.org/content/75/14/e59.full.html##ref-list-1

Citations
This article has been cited by 1 HighWire-hosted articles:
http://www.neurology.org/content/75/14/e59.full.html##otherarticles

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
All Clinical Neurology
http://www.neurology.org/cgi/collection/all_clinical_neurology
All Spinal Cord
http://www.neurology.org/cgi/collection/all_spinal_cord
Diplopia (double vision)
http://www.neurology.org/cgi/collection/diplopia_doubleVision
MRI
http://www.neurology.org/cgi/collection/mri
Primary brain tumor
http://www.neurology.org/cgi/collection/primary_brain_tumor

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/misc/about.xhtml#permissions

Reprints
Information about ordering reprints can be found online:
http://www.neurology.org/misc/addir.xhtml#reprintsus