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SYLVIUS AQUEDUCT SEPTUM

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We present a case of chronic hydrocephalus discovered in adulthood through an episode of acute decompensation. Multimodal imaging revealed the cause of this hydrocephalus to be a membranous septum of the aqueduct of Sylvius, a condition for which few reports exist.

Key-word: Brain, hydrocephalus.

Case report

A 42-year-old man with no significant past medical history was admitted to the emergency room with a complaint of severe headache and nausea for the last 24 hours. Lower limbs weakness was noted upon physical examination.

A non-contrast followed by a contrast-enhanced computed tomography (CT) examination of the head was performed (Fig. 1 and 2) and revealed severely dilated lateral and third ventricles, with an Evans' index estimated at 0.42, effacement of cortical sulci over the convexity and periventricular halo-shaped hypodensity. Sylvius aqueduct was found to be also enlarged, but the fourth ventricle appeared within normal limits. No mass or other density abnormality was seen.

These findings were in keeping with chronic hydrocephalus related to the major enlargement of the lateral and third ventricles and signs of cerebrospinal fluid (CSF) transependymal resorption, in acute decompensation, presenting with intracranial hypertension symptoms.

The patient underwent emergent right ventricular catheterization with a subcutaneous Rickham's reservoir as to relieve the acute symptoms.

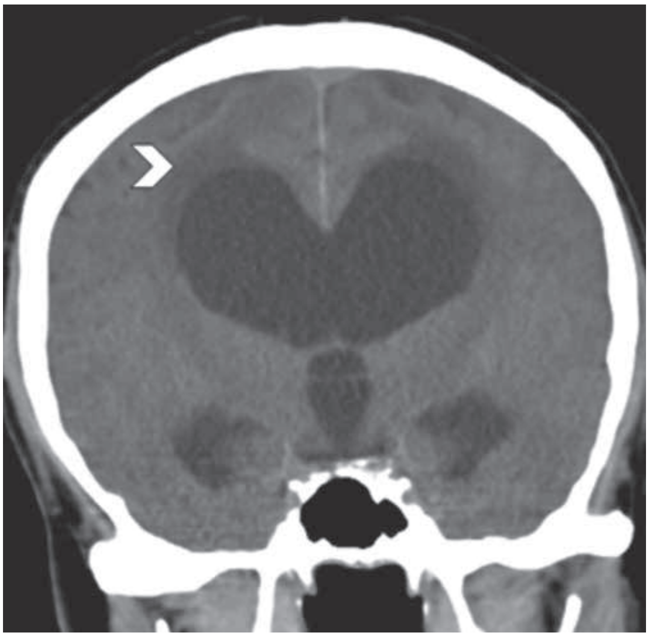

Fig. 1. — Non contrast-enhanced head CT multiplanar reformation (MPR) coronal slice displaying severely enlarged lateral ventricles and periventricular hypodensities (arrowhead) compatible with transependymal CSF resorption.

Etiologic workup performed a few days later comprised a CT ventriculography with injection of a water-soluble contrast medium (Omnipaque 240, GE Healthcare) through the Rickham's reservoir (Fig. 3 and 4) and a magnetic resonance imaging (MRI) including time-of-flight, gradient echo and flow-sensitive sequences (Fig. 5 and 6).

It revealed a Sylvius aqueduct stenosis by a membrane-like obstacle at its junction with the 4th ventricle. The tectal plate is displaced posteriorly, as typically seen in distal aqueduct stenosis (1).

Moreover, these post-surgical head examinations showed significant re-enlargement of cortical sulci and decreased third ventricle floor bulging, accounting for diminished intracranial pressure and treatment success.

Opacification of the 4th ventricle during ventriculography demonstrates permeability of the membranous obstacle, at least when ventricles are in a less dilated state.

MRI ruled out other local abnormalities; especially there was no evidence of vascular pathologies, tumors or signs of micro-bleeds. CSF flow through the Sylvius aqueduct after ventricular derivation is shown to be non hyperdynamic as suggested by the absence of flow void on the sagittal T2w images and the absence of flow intensity on the flow-sensitive sequence.

The patient fared well and underwent an endoscopic third ventriculocysternostomy a few weeks later with no early complications.

Discussion

Sylvius aqueduct is the most common site of blockade of CSF flow through the ventricles (1).

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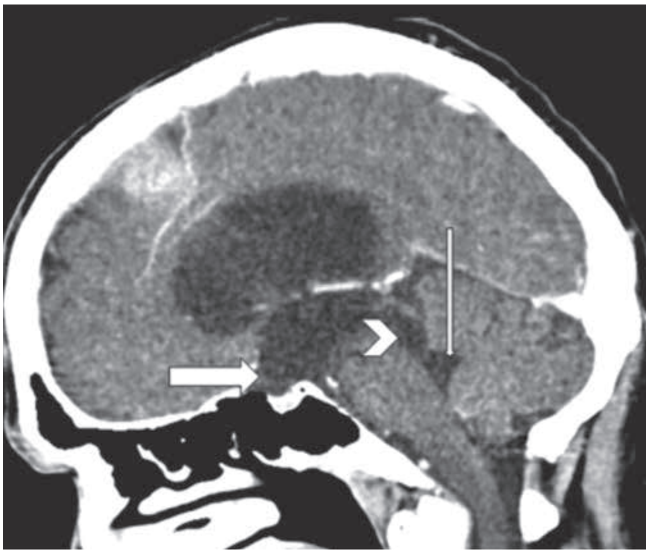

Fig. 2. — Contrast-enhanced head CT MPR median slice showing dilated aqueduct of Sylvius (arrowhead) and third ventricle with bulging of its floor (thick arrow), but normal appearing fourth ventricle (thin arrow). The subarachnoid spaces over the convexity are narrowed.

Fig. 4. — CT ventriculography volume rendering technique after stripping of the skull structures showing the stenosis of the aqueduct (arrow).

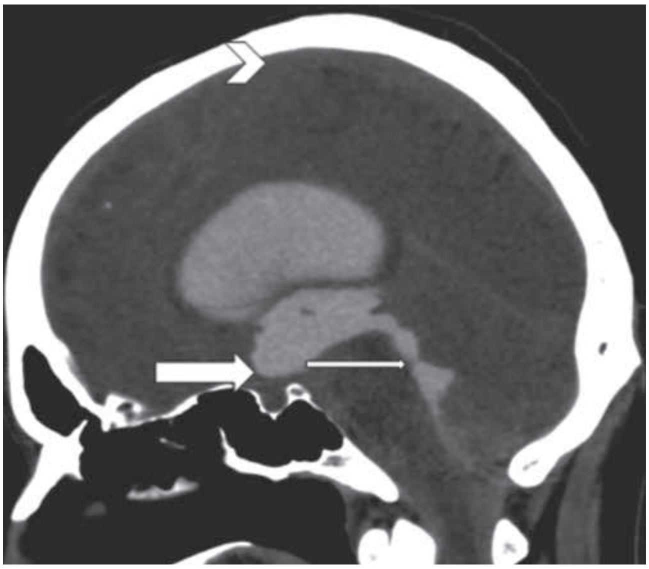

Fig. 3. — CT ventriculography MPR median slice showing decreased third ventricle floor bulging (thick arrow) and increased subarachnoid space over the convexity (arrowhead), a stenosis by a membrane-like structure in the distal portion of the Sylvius aqueduct (thin arrow), and opacification of the 4th ventricle, cisterna and subarachnoid spaces over the convexity.

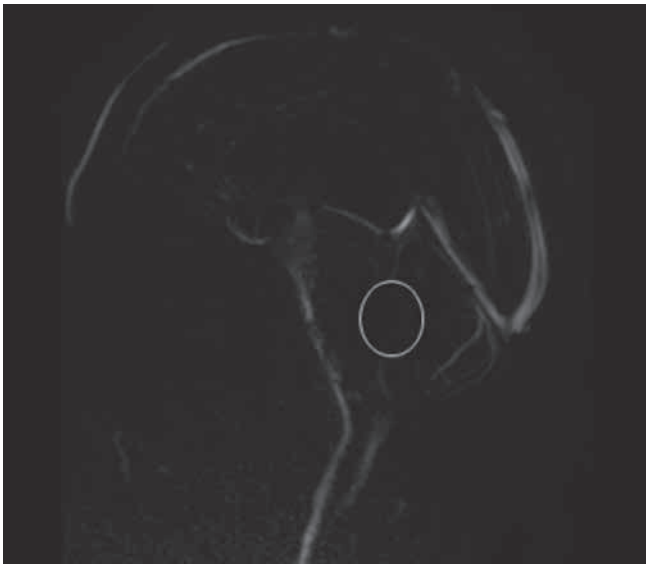

Fig. 5. — Sagittal maximum intensity projection of a flow-sensitive MR sequence tuned to CSF velocities displaying the absence of flow in the aqueduct of Sylvius (circle).

Presentations of benign aqueduct stenosis in adulthood is not infrequent (2) and constitutes the first cause of obstructive hydrocephalus in adults (3).

Differential diagnosis may be classified into primary and secondary causes. Secondary causes encompass extrinsic space occupying lesions while primary causes are intrinsic lesions of the aqueduct and have been classified by Russell into four types (1): stenosis (normal aqueduct cells within one abnormally narrow aqueduct), forking (division into two or more channels), gliosis (reaction to injuries such as infection, hemorrhage or toxins) and membranous occlusion.

The latter is usually distally located and is formed by a gliotic membrane.

Our patient had no history of congenital infection, meningitis or subarachnoid hemorrhage. MRI

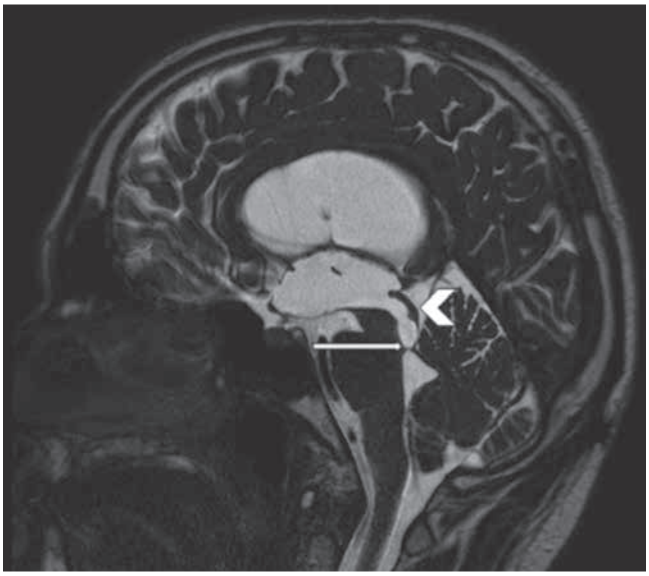

Fig. 6. — Sagittal T2-weighted SE MR slice confirms the membrane-like obstacle in the caudal part of the aqueduct of Sylvius (arrow). The tectal plate is displaced posteriorly (arrowhead) in this distal aqueduct stenosis.

Note the absence of the normal CSF flow void in the aqueduct.

exploration revealed no signs of neoplastic lesions or remote bleeding. The membranous obstacle in the distal part of the aqueduct was clearly demonstrated on the CT ventriculography and the T2W sagittal sequence. This lesion is likely to be developmental and idiopathic in its nature.

Few cases of such membranous occlusions have been reported in the late seventies (4, 6).

Mechanisms of compensation to increased CSF pressure upstream to the obstacle in chronic obstructive hydrocephalus, e.g. increased periventricular capillary CSF resorption, account for many phenomena in the physiopathology (7). In this setting, ventricular expansion is allowed under near normal intracranial pressure and cerebral perfusion is diminished.

Sylvius aqueduct stenosis is not a stable condition (1). Functional processes such as deformation of the brain stem due to the ongoing enlargement of the supratentorial ventricles may worsen the obstacle and precipitate acute intracranial hypertension.

Opacification of the 4th ventricle and subarachnoid spaces in the postdrainage CT ventriculography study pleads for residual permeability through the membrane, at least when the ventricles are in a decompressive state.

Conclusion

We presented a case of chronic hydrocephalus discovered in adulthood through an episode of acute decompensation. Multimodal imaging revealed the cause of this hydrocephalus to be a membranous septum of the aqueduct of Sylvius, a condition for which few reports exist.

We think that partial permeability of the membrane and compensation mechanisms explain the late presentation of the patient, with no previous obvious symptoms. The acute episode is likely to be due to functional worsening of the obstacle by ventricular deformation.

References

1. Cinalli G., Spennato P., Nastro A., Aliberti F., Trischitta V., Ruggiero C., et al.: Hydrocephalus in aqueductal stenosis. Child's nervous system: ChNS: official journal of the International Society for *Pediatric Neurosurgery*, 2011, 27: 1621-1642.
2. Harrison M., Robert C., Uttley D.: Benign aqueduct stenosis in adults. *J Neurol, Neurosurg & Psych*, 1974, 37: 1322-1328.
3. Tisell M.: How should primary aqueductal stenosis in adults be treated? A review. *Acta Neurol Scand*, 2005, 111: 145-153.
4. Gökalp H., Tascioglu A.: Membranous occlusion of the aqueduct of Sylvius. *Surg Neurol*, 1977, 8: 103.
5. Murphy M.J., Lyon L.W., Carlstrom T.A., McDonnell D.E.: Normal pressure hydrocephalus due to membranous obstruction of the sylvian aqueduct. *Neurology*, 1978, 28: 835.
6. Ludwiczak R., Dietemann J., Wackenheim A.: CT and pneumographic studies of membranous occlusion of the aqueduct of Sylvius. A case report. *Neuroradiology*, 1979, 18: 53-55.
7. Egnor M.: Radiological assessment of hydrocephalus: new theories and implications for therapy. *Neurosurgical Review*, 2004, 27 (3).