Blake's pouch cyst

Blake's pouch cyst is a cystic appearing structure that represents posterior ballooning of the inferior medullary velum into the cisterna magna, below and posterior to the vermis that communicates with an open fourth ventricle. It is caused by a failure of regression of Blake's pouch secondary to the non-perforation of the foramen of Magendie.

History and etymology

It is named after Joseph A. Blake (1864-1937), an American physician who studied the physiology related to the foramen of Magendie

Blake's pouch cyst was first described as an independent entity within the Dandy-Walker malformation in 1996 by Paolo Tortori-Donati et al.

Epidemiology

It is a rare and underdiagnosed entity.

Pathology

Blake's pouch, also known as the rudimental fourth ventricular tela choroidea, is a normal transient structure during embryological development which regresses, usually by 12 weeks of gestation, when it starts fenestrating to form the foramen of Magendie (which forms in up to 4th month of gestation).

Persistent Blake's pouch cysts occur due a failed perforation of the foramen of Magendie. As the foramina of Luschka open later than the foramen of Magendie during the embryologic development, this non perforation of the foramen of Magendie causes enlargement of the ventricular system until the foramina of Luschka open and establishes a precarious compensation with CSF flowing into the cisterns.

Blake's pouch cysts used to be classified as part of the Dandy-Walker continuum, however, this entity is not related to cerebellar or 4th ventricle malformations

Clinical presentation

It may be asymptomatic or it can present:

in children or in adulthood with signs of hydrocephalus (usually headache, vomiting, blurred or double vision)

later in life or be even asymptomatic, probably due to adequate CSF flow through lateral foramen of Luschka

Diagnosis

Radiographic features

MRI

MRI is the imaging modality of choice due its capacity to differentiate this entity from other posterior fossa malformations. Features include:

infra vermian cyst that communicates with fourth ventricle

cyst is smooth with thin walls: it can be visualised on thin sagittal T2 images

it can impress on medial side of cerebellar tonsils due to size

cyst does not communicate with the cisterna magna posteriorly

no vermian hypoplasia or rotation

usually hydrocephalus of 4th and supratentorial ventricles (tetraventricular hydrocephalus)

elevation of the tentorium but usually normally positioned torcular

choroid plexus can extend from 4th ventricle into superior portion of cyst, which is essentially a ventricular diverticulum.

Differential diagnosis

It needs to be distinguished from other causes of enlarged retrocerebellar “CSF” space:

mega cisterna magna: communicates freely with both 4th ventricle and subarachnoid space

other entities of the Dandy-Walker continuum: cerebellar vermis malformations

arachnoid cyst: no hydrocephalus

Treatment

Preferable treatment of hydrocephalus is endoscopic third ventriculostomy. The prognosis is good after treatment.