Neuroenteric cyst of the cerebellum

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Summary

Intracranial neuroenteric cysts are rarely encountered and no more than 13 cases have been reported in the literature. This paper reports another case located in the cerebellum. These occur mostly in adults and there is no typical clinical presentation. The computed tomographic scan shows them as low attenuating lesions with no peripheral contrast enhancement. These cysts are believed to be developmental in origin.

KEY WORDS: headache, optic atrophy.

Introduction

Neuroenteric cysts (synonyms: enterogenous cysts, neuroepithelial cysts, gastrocytoma) are usually defined as cysts lined by a single layer of mucin containing columnar or cuboidal epithelium which are generally non-ciliated. These have been mostly reported in the spinal canal either in the dorsal or lower cervical region (Hirano *et al.*, 1971). Their occurrence in the posterior fossa is rare and no more than 13 cases have been reported in the literature (see below).

This paper reports one more case which was located in the vermis of the cerebellum. The controversies of its origin and the differential diagnosis are discussed.

Case report

A 48-year-old man was admitted in February 1980 with occipital headaches for 2.5 years and deteriorating vision, giddiness and unsteadiness of gait for 3.5 months. Examination revealed minimal proptosis of the left eye and a café-au-lait patch on the right chest wall. He was conscious and oriented but had no vision in the right eye and only perception of hand movement in the left eye. Fundus showed bilateral

secondary optic atrophy. There was no other cranial nerve deficit. The deep tendon reflexes on the right were brisker and the plantar response was extensor. No motor weakness or sensory loss was detected, and except for mild unsteadiness of gait no other cerebellar signs could be elicited.

X-rays of the skull showed rarefaction of the dorsum sellae. A left carotid angiogram suggested dilated lateral ventricles. Computed tomography with contrast enhancement showed a moderate degree of hydrocephalus with a well-defined round low attenuating lesion in the region of the 4th ventricle with no peripheral enhancement (Fig. 1). An air cum Amipaque® ventriculogram showed a filling defect in right lateral recess of the 4th ventricle and an upward displacement of the inferior medullary velum (Fig. 2).

A ventriculo-peritoneal shunt relieved his headache. A fortnight later a midline sub-occipital

FIG. 1. Computed tomogram showing well-defined low attenuating lesion in the region of the fourth ventricle. No enhancement occurred with administration of contrast material.

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FIG. 2. Lateral view of Amipaque ventriculogram showing an upward displacement of the inferior medullary velum.

craniotomy was performed. The vermis appeared broad and on incising it a sub-cortical white cyst was seen. Careful dissection revealed a large oval cyst (3×2 cm) containing thick gelatinous fluid which was delivered with ease. The post-operative period was uneventful and the patient was discharged on the 12th day with no headaches but with mild unsteadiness of gait.

Microscopically, the cyst was lined by a single layer of columnar epithelium. The cells had round basally situated nuclei, with clear vacuolated cyto-

plasm, and no cilia were identified. No pleomorphism or mitosis was seen. The epithelium rested on an outer coat of collagen and the lining cells contained mucin which was confirmed by mucicarmine stain.

Discussion

The common non-neoplastic cystic lesions encountered in the posterior fossa include the Dandy-Walker cyst, an arachnoidal cyst, and a large cisterna magna. An epithelial lined cyst in the posterior fossa is seldom considered in the differential diagnosis. Dandy-Walker cysts are common in infants with hydrocephalus and arachnoidal cysts are encountered in somewhat older children and only occasionally in adults (Matson, 1969). In contrast, all the posterior fossa neuroepithelial cysts reported in the literature have been in adults (Table 1); the youngest reported patient was 22 years of age and the oldest 68 years, with a striking male preponderance (9 males to 5 females). These cysts are generally located in the midline but may also lie in the cerebello-pontine angle. Details of the clinical presentation are available only in 10 cases and among them two were asymptomatic. The duration of symptoms ranged from 2 months to 4 years with four of the eight cases having symptoms for less than 1 year. The most frequent complaint was nuchal pain (seven cases), Papilloedema and impairment of vision was less frequent and was present in only three cases. Symptoms and signs of cerebellar involvement were

TABLE 1. Details of posterior fossa neuroepithelial cysts reported in the literature

Author	Location	Age Sex	Findings	Histology	PAS stain	Result
Dandy, 1969	CPA*	Adult	—	—	—	—
Parkinson and Childe, 1952	4th ventricle	28 M	Two cysts, gelatinous fluid. Excised	Columnar ciliated epithelium	?	Improved
Shuangshoti and Netsky, 1966	4th ventricle	48 M	Pedunculated single cyst, 8 mm diameter	Simple cuboidal	+	Autopsy
Shuangshoti and Netsky, 1966	4th ventricle	60 M	Cyst, 4 mm diameter	Simple cuboidal	?	Autopsy
Shuangshoti and Netsky, 1966	4th ventricle	50 M	Cyst, 2 mm diameter	Simple cuboidal	+	Autopsy
Hoening <i>et al.</i> , 1967	4th ventricle	68 M	Multiple cysts mucoid material	Low cuboidal/columnar epithelium	+	Autopsy
Afshar and Scholtz, 1981	4th ventricle	48 M	Clear colourless fluid excised	Non-ciliated cuboidal epithelium	+	Improved
Small, 1962	Prepontine	—	—	—	—	?
Small, 1962	Prepontine	—	—	—	—	?
Small, 1962	Prepontine	—	—	—	—	?
Hirai <i>et al.</i> , 1981	Prepontine	30 F	Turbid fluid. Excised	Cuboidal or columnar epithelium Occ. cilia	+	Improved
Giombini and Lodrini, 1981	Vermis	37 M	Xanthochromic fluid	Columnar or cuboidal epithelium	+	Improved
Hasegawa <i>et al.</i> , 1966	Vermis	22 M	Clear fluid. Excised	Ciliated non-ciliated cuboidal epithelium	?	Improved
Present case, 1982	Vermis	48 M	Thick gelatinous fluid, Excised	Non-ciliated columnar epithelium	+	Improved

*Cerebello-pontine angle.

present in the majority. The involvement of the 5th nerve in two patients is not easily explicable, except by direct pressure. One patient had bilateral sensory neural deafness which was attributed to raised intracranial pressure (Saxena *et al.*, 1969).

The fluid in these cysts was either clear, gelatinous, mucoid or xanthochromic. In seven cases the cells of the cyst wall contained P.A.S. positive material within the cytoplasm. In three cases the authors fail to mention the details about the staining.

It is usually difficult to diagnose these lesions preoperatively even after ventriculography. Computed tomography shows these cysts as clearly defined round areas of low attenuation but with no peripheral contrast enhancement. The 4th ventricle may not be clearly made out as in our case. Associated hydrocephalus was present in four of the 10 reported cases. Cystic astrocytoma and heman-gioblastoma may be confused with these lesions but these generally contain areas of increased attenuation within them.

The origin of these cysts has been a matter of some debate. They have been considered as a form of developmental anomaly because of an incomplete separation of the primitive gut from the neural groove. The enterogenous cysts of the spinal canal are usually associated with bony anomalies of the vertebrae (Beardmore and Winglesworth, 1958; Bremer, 1952; McLetchie *et al.*, 1954), but these may occasionally be absent (Fabinyi and Adams, 1979; Guilburd, Ben Arie and Peyser, 1980; Scoville *et al.*, 1963). Their origin from ectopic ependymal cells has also been postulated; a part of the ventricular ependymal lining becoming pinched off and sequestered during embryogenesis (Shuangshoti and Netsky, 1966). Sequestered ependymal cells have been seen well away from the lining of the ventricles as in the occipital lobes (Cooper and Kernohan, 1951).

Phospho-tungstic acid haematoxylin stain failed to show any glial fibres in the wall of the present case to suggest a possible glial origin. The lining in our case was non-ciliated and mucicarmine positive which is unlike an ependymal or an arachnoidal cysts. We concur with the view expressed by Giombini and Lodrini (1981) that these cyst originate from the most cranial portion of the primitive intestine.

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