Foramen Magnum Neurenteric Cyst Causing Mollaret Meningitis: MR Findings

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Summary: MR showed a neurenteric cyst posterior to the cervicomedullary junction in a man with chronic recurrent aseptic meningitis. On T1-weighted MR, the cyst was of high intensity in relation to the brain, and on T2-weighted MR it was of less intensity than cerebrospinal fluid.

Index terms: Brain, cysts; Meningitis; Foramina, magnum

Chronic, recurrent, aseptic meningitis with polymorphonuclear pleocytosis and negative cerebrospinal fluid (CSF) cultures (Mollaret meningitis) has been reported secondary to subarachnoid leakage of fluid, usually from epidermoid cysts (1, 2). We report a case of Mollaret's meningitis caused by leakage from a foramen magnum neurenteric cyst.

Case Report

A 32-year-old man presented with fever, headache, nausea, vomiting, photophobia, a "sore neck," and a 6-month history of intermittent dull headache. Physical examination revealed fever, moderate meningismus, and positive Kernig's and Brudzinski's signs. Lumbar puncture revealed pleocytosis (white blood cell count, 4400/mm^3, 100% polymorphonuclear), elevated protein (123 mg/dL), and low glucose (8 mg/dL). He was admitted to his local hospital, and ceftriaxone and vancomycin were given for presumed bacterial meningitis. However, CSF and blood cultures were negative, and he did not improve after 3 days of treatment.

Brain computed tomography without contrast was interpreted as normal but did not include the entire foramen magnum. Subsequent lumbar punctures showed variable amounts of polymorphonuclear cells and monocytes, the latter of which were often atypical. No microorganisms were seen, and subsequent blood and CSF cultures, including studies for fungi and acid-fast bacilli, were negative. The patient was empirically given fluconazole for putative *Candida* meningitis. He gradually improved, and subsequent lumbar puncture demonstrated a significantly reduced white blood cell count. He was discharged and received oral fluconazole.

Four weeks later he returned with recurrent symptoms. CSF culture revealed a white blood cell count of 1930/mm^3, 97% polymorphonuclear cells and 3% monocytes. He was transferred to our institution for further evaluation

Brain and whole-spine magnetic resonance (MR) demonstrated a lobulated, nearly homogeneous-intensity mass in the posterior aspect of the foramen magnum extending within the cervical subarachnoid space down to the C-1 level (Fig 1A–C). Compared with CSF and the cord, it was of markedly increased signal intensity on noncontrast T1-weighted (566/11/2 [repetition time/echo time/excitations]) images. On T2-weighted (4000/90/1) images, the lesion was hypointense compared with CSF and slightly hyperintense compared with cord. There was mild distortion of the cerebellum and cervicomedullary junction. The posterior basiocciput appeared to be slightly remodeled by the mass, but there was no bone invasion. There was minimal contrast enhancement of the outer layer of the mass.

Suboccipital craniectomy and cervical laminectomy were performed with removal, and fixation, of the intact cyst. The resected cyst had a white, firm wall with an inner epithelial lining, which surrounded brown, nearly solid, material. Microscopically, the cyst contained macrophages, degenerating epithelial cells, and keratin. The majority of the epithelium was columnar and cuboidal (Fig 1D). The epithelium was one to two cell layers thick, the basal layer being predominantly cuboidal. The epithelial surfaces varied from ciliated, to microvillous, to flat. There were a few regions of simple squamous epithelium (Fig 1E). The epithelium lined a wall of highly fibrillar collagen. Portions of the mass were infiltrated with macrophages filled with blue-gray granular material that resembled the dead cellular debris and keratin contained within the cyst (Fig 1D). Some portions of the cyst wall were thinned and chronically inflamed, with macrophages outside of the cyst wall, suggesting prior leakage.

The patient has been followed for 2 years since this resection with no further episodes of meningitis.

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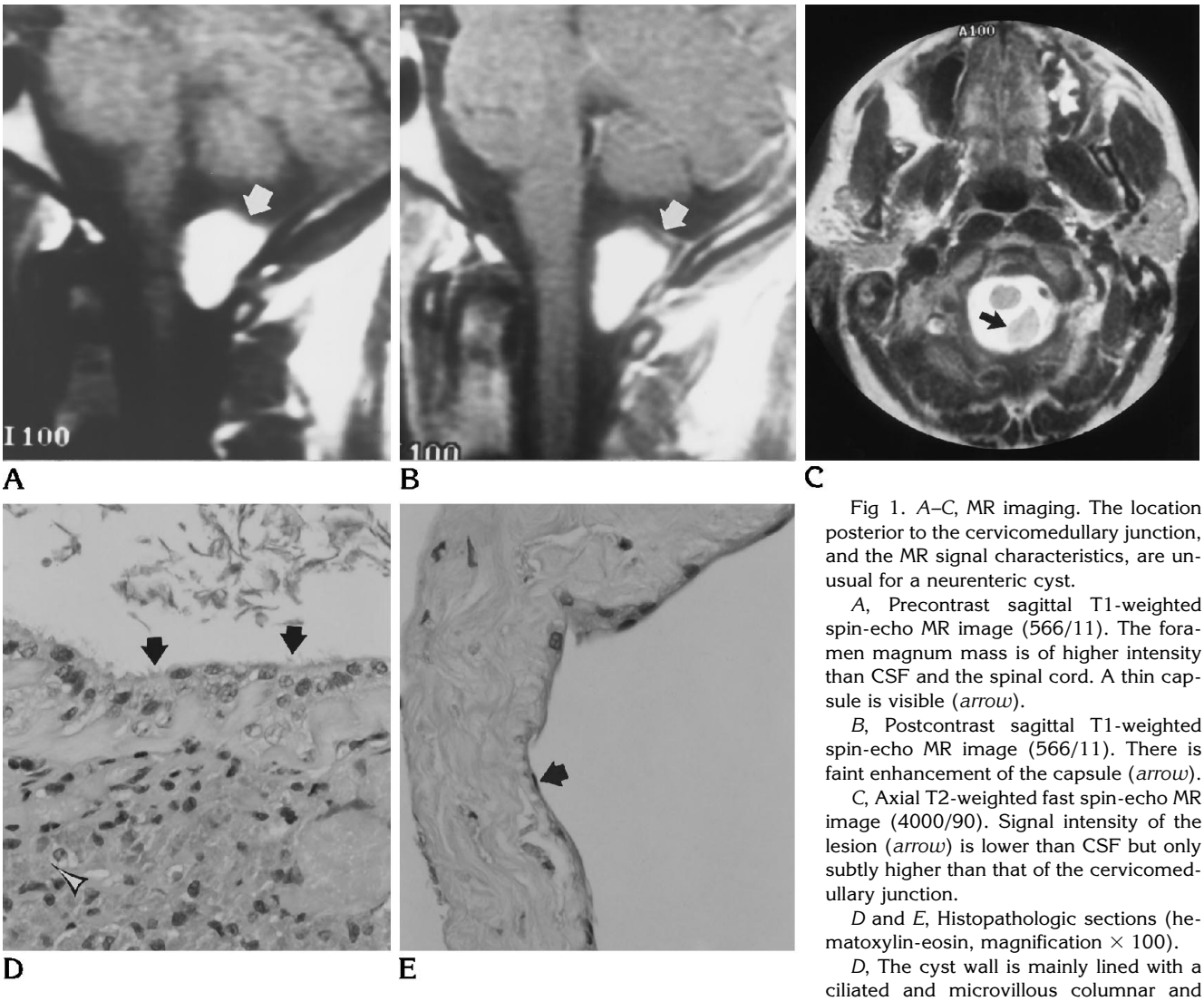

Fig 1. *A–C*, MR imaging. The location posterior to the cervicomedullary junction, and the MR signal characteristics, are unusual for a neurenteric cyst.

A, Precontrast sagittal T1-weighted spin-echo MR image (566/11). The foramen magnum mass is of higher intensity than CSF and the spinal cord. A thin capsule is visible (*arrow*).

B, Postcontrast sagittal T1-weighted spin-echo MR image (566/11). There is faint enhancement of the capsule (*arrow*).

C, Axial T2-weighted fast spin-echo MR image (4000/90). Signal intensity of the lesion (*arrow*) is lower than CSF but only subtly higher than that of the cervicomedullary junction.

D and *E*, Histopathologic sections (hematoxylin-eosin, magnification × 100).

D, The cyst wall is mainly lined with a ciliated and microvillous columnar and cuboidal epithelium (*arrows*), favoring an endodermal origin. Macrophages (*arrowhead*) infiltrating the wall contain amorphous phagocytosed material that resembles the keratin and dead cellular debris that fill the cyst lumen.

E, A few portions of the cyst wall are lined with a simple squamous epithelium (*arrow*). Chronic pressure effects can induce the columnar and cuboidal epithelium to resemble a squamous epithelium.

Discussion

Mollaret meningitis is a recurrent aseptic meningitis associated with polymorphonuclear-dominant CSF pleocytosis. Usually, there is also monocytosis with variably atypical cells. If they are markedly atypical, they are known as Mollaret cells (1–3). Although the cause of Mollaret meningitis is believed to be viral, two reports of epidermoid cysts causing Mollaret meningitis suggested that leakage from the cyst results in chemical meningitis (1, 2). Although Mollaret cells were not identified in our case, the dominant polymorphonuclear pleocytosis, the variable, atypical, monocytosis, and negative CSF cultures are all consistent with Mollaret meningitis.

The unusual location and mixture of epithelial elements in our cyst somewhat confounds precise pathologic classification. However, the predominance of a ciliated and microvillous pseudostratified columnar and cuboidal epithelium argues for an endodermal origin (4). Neurenteric cysts have been reported in the posterior fossa, including the foramen magnum (5–8). However, such cysts are more common in the cervical and upper thoracic meninges (9), where they are usually anterior to the cord, associated with vertebral defects, and present with

spinal cord compression (10). Similar cysts can also occur in the lumbar spine (11).

The cytogenesis of the simple squamous epithelium in our cyst is uncertain. Cysts with mixed epithelium suggest either transition from arachnoidal cells to respiratory epithelium, or flattening of the epithelial cells from expansion of the cyst (12). We believe that it is most likely that the squamous epithelium in our case resulted from pressure caused by the cyst contents. Local regions of squamous cell metaplasia probably provided the keratin within the cyst contents. There was local wall thinning and inflammation, consistent with prior leakage. Macrophages found outside the wall of the cyst contained phagocytosed debris resembling cyst contents, further supporting prior leakage.

Epidermoid cysts causing Mollaret meningitis have been reported (1–3); leakage from neurenteric cysts is a rare cause of aseptic meningitis (11, 13). A review of MR imaging of neurenteric cysts has described their T1-weighted signal intensity as usually lower than that of the cord and equal to, or slightly higher than, that of CSF (8). The T2-weighted signal was reported usually to be higher than that of the cord but equal to, or slightly higher than, that of CSF. Although our pattern of signal changes is, therefore, unusual for neurenteric cysts, embryonic cysts can have many variations in protein content, hemorrhage, cholesterol content, and other materials (8). This would be especially true of lesions with mixed histology, such as ours. Portions of our lesion, if viewed without knowledge of the appearance of the other portions, could have been classified as coming from an epidermoid cyst. The location of our lesion posterior to the cervicomedullary junction, and the lack of neuraxis anomalies, are more commonly seen in epidermoid cysts but have been reported in neurenteric cysts (8).

Thorough imaging of the neuraxis is required in patients with clinical findings suggesting Mollaret meningitis. If the intracranial contents show no lesion, the spinal neuraxis needs to be searched.

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