A 13-year-old boy presented to the neurosurgical services with complaints of bifrontal headache for 1 year and progressive deterioration of vision in both eyes. The headache had worsened for the past 1 month, associated with vomiting at the height of headache. Fifteen days prior to admission to the hospital his visual acuity worsened to an extent just to perceive light. General systemic examination was normal. The neurological evaluation revealed visual acuity of only perception of light, bilateral secondary optic atrophy, right sixth cranial nerve paresis, and left cerebellar signs. On cranial CT scanning an oval, hypodense (Figure 1A) left cerebellar cystic mass was noted, nonenhancing with contrast (Figure 1B). Fourth ventricle was compressed with resultant hydrocephalus. With a clinical diagnosis of cystic cerebellar tumor, the cyst was excised by suboccipital craniectomy. The cyst ruptured during the surgery and was found to be lined by thin white membrane and filled with xanthochromic fluid.

Pathological examination

The resected material had a cystic zone with cerebellar folia adherent to it on the external surface. Microscopic examination of the cystic lesion revealed compressed cerebellar white matter lined by foamy histiocytes and reactive astrocytosis (Figure 2). The cerebellar folia had features of longstanding compression and the overlying meninges were markedly thickened. There was no mural nodule or a glioma. Internal to the histiocytic layer a thick multilamellar eosinophilic, chitinous wall and a syncitial germinal layer was present. The lumen had proteinaceous granular material admixed with erythrocytes and numerous brood capsules of *Echinococcus granulosus* in varying stages of evolution (Figure 3). Many oval to round calcific concretions were seen probably representing the degenerated parasitic ova.

Diagnosis: Hydatid cyst in cerebellum

Discussion

*E. granulosus* (Hydatidosis) infestation is a zoonotic disease, humans getting infected accidentally as intermediate hosts. The disease is transmitted usually by food-borne contamination by eggs passed in faeces of definitive host, mainly dogs. *E. granulosus* infestation results in liver cysts in nearly half the cases and brain involvement is less than 5%. The symptomatology, depends crucially on the location of the cyst in the brain.[1] The infestation may be primary often presenting as solitary parenchymal cyst. Secondary involvement is due to cyst rupture or hydatid fluid dissemination from a distant organ, by haematogenous route. CNS echinococcosis is rare in infants and more common in children up to 15 year of age than in adults. The signs and symptoms include raised intracranial pressure, headache, vomiting, papilloedema, hemiparesis and occasionally seizures.[1] The cyst lodges frequently in the cerebral hemisphere, attaining a large size. Involvement of the cerebellar hemisphere and presenting radiologically as a cystic mass, is usually mistaken for cerebellar

![Figure 1: (A) Plain CT scan showing large, oval, hypodense left cerebellar cystic mass. (B) Contrast enhanced CT showing nonenhancing, hypodense left cerebellar cystic mass](image-url)
Figure 2: Low magnification view of cerebellar lesion showing compressed cerebellar white matter lined by foamy histiocytes and reactive astrocytes

Figure 3: The lesion has a thick lamellar, chitinous wall, a germinal layer and numerous brood capsules of *E. granulosus*. The lesion has a germinal layer and numerous brood capsules of *E. granulosus*

astrocytoma as in the present case. Slight or lack of ring enhancement or marginal edema on MRI should arouse suspicion of hydatidosis, especially in paediatric age patients from the tropics.[2] Recently *in vivo* proton magnetic resonance spectroscopy has been able to identify intra-axial hydatid and other parasitic cysts from other cystic lesions.[3,4] Rupture of the cyst during surgery may trigger anaphylaxis followed by disseminated infection.[5] At our center, a female child was operated on thrice to resect the cysts and each time a new crop appeared again. Laboratory diagnosis by ELISA, immunoblot and indirect immunofluorescence to detect the antigen and immune complexes assist in diagnosis. Negative serological tests do not rule out the disease. Live cyst elicits negligible host reaction and surgical specimens rarely include brain tissue. The cyst undergoing degeneration, elicits epitheloid and giant cell reaction, finally leaving behind amorphous necrotic debris surrounded by dense gliosis. Following infestation by *E. parasite*, the host develops an immunological response protective against reinfection, but not effective against the lodged parasite. Evasion of the host immune attack against the parasite is mediated by the suppression of T-lymphocyte function and inhibiting macrophage-lymphocyte interaction.[6] Persistent production of IL-4 as a result of chronic infection could lead to enhanced production of IgG4 antibodies, which probably act as blocking antibodies, binding with lipoprotein antigen B in hydatid cyst, thus down regulating the host response and protecting against anaphylaxis.[7]

References


Accepted on 20-07-2005