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Abstract: Intracranial epidermoid tumors are rare (0.2–1.8%), histologically benign, slow-growing, congenital neoplasms of the central nervous system arising from the retained ectodermal implants. We report an unusual case of intra third ventricular epidermoid in an adult presenting with the Bobble-head doll syndrome and review the relevant literature.

Key words: epidermoid tumor, third ventricle, Bobble head doll syndrome

Introduction

Intracranial epidermoid tumors are rare (0.2–1.8%), histologically benign, slow-growing, congenital neoplasms of the central nervous system arising from the retained ectodermal implants. (1-4) These lesions tend to spread along normal cleavage planes, and slowly fill any available subarachnoid space, including the sulci, cisterns, fissures, and ventricles. (4, 5) Because of this unique behavior the expanding tumor conforms to the shape of the cavities, however the tumor does adhere to and surround the vital structures. (6, 7)

Case Reports

A 55 year gentleman presented with headache of 6 months duration with vomiting off and on for last 15 days. He also had Bobbing movements of the head. There was no

history of focal motor or sensory weakness. His general and systemic examination was unremarkable. Higher mental functions were normal. Cranial nerves were normal except bilateral papilloedema. Extraocular movements were and there was no nystagmus. His voice was of low volume and without slurring. Motor power was normal and sensory functions were also normal. He was able to walk with support. He had fine tremors in both the hands. On examination he had bobbing movements of the head (Movie). Computed tomography scan (CT scan) of the brain showed a large discrete area of radiolucency in the third ventricle and there was ventriculomegaly without any evidence of periventricular edema. (Figure 1). Magnetic resonance (MR) imaging of the brain was showed a large, mass lesion in the third ventricle also insinuating in the basal cisterns;

however there was no extension of the lesion into the fourth ventricle (Figure 2). The mass was slightly hyperintense relative to CSF on T1-weighted images, hyperintense T2-weighted with heterogeneous appearance and heterogeneous nature was better appreciated on FLAIR images. On diffusion-weighted (DW) images, there was overall restricted diffusion with areas of free diffusion. There was no significant enhancement of the tumor after intravenous administration of gadolinium contrast material. Based on these imaging findings a diagnosis of intraventricular epidermoids was made. (8-11) The patient underwent right parietal parasagittal craniotomy, interhemispheric transcallosal approach and near total decompression of the tumor. After opening the splenium of the corpus callosum a pearly structure with an appearance characteristic of an epidermoid tumor was seen filling the ventricle completely, that was near totally decompressed. Histopathological evaluation showed typical features consistent with epidermoid. The patient had unremarkable postoperative course and improved in his abnormal movements and gait.

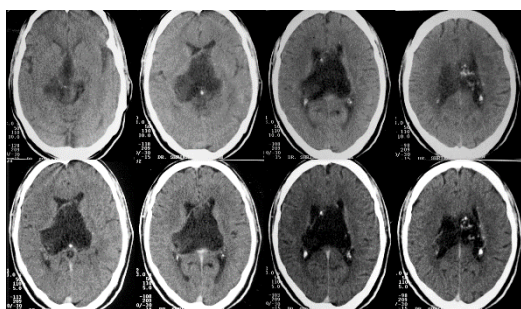


Figure 1 - CT scan showing large third ventricular epidermoid

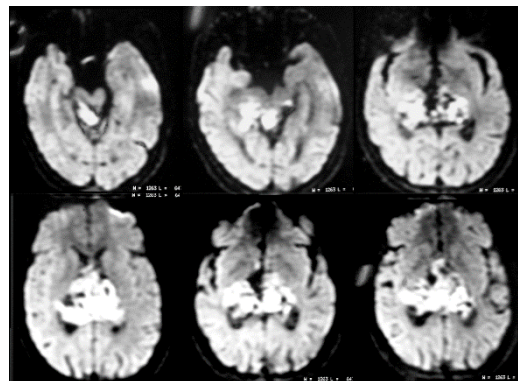


Figure 2 - MRI showing extension of the lesion

Discussion

The commonest sites of intracranial epidermoids include cerebellopontine angle and the chiasmal region, however these lesions can also appear in the cerebral hemisphere and the intraventricular cavities. (2, 8, 12) Intraventricular location of the particularly third and lateral ventricle is extremely uncommon with only few cases in the literature. (13-16) They can be a direct occurrence of the epidermoids in the ventricular or the ventricles can be secondarily involved as in present case, (16-18) however it is often extremely difficult to say exactly where an epidermoid has originated. (17-19) The Bobble-head doll syndrome is a rare and unique movement disorder characterized by continuous or episodic involuntary forward and backward and side to side movement of the head that mainly encountered in children. (20-22) The etiology of this syndrome include third ventricular tumors, suprasellar arachnoid cysts, aqueductal stenosis and other lesions in the region of the third ventricle along with communicating hydrocephalus. (20, 22) Usually the decompression of the

third ventricle results in the abolition of the abnormal movements. (20, 22) As in present case these movements can be abolished after the removal of the ventricular lesion. (8) Although total removal is ideal, but close proximity of tumours to cranial nerves and the brain stem pose technical difficulties in total removal. (9, 10) Endoscopic removal is another option for the removal of these tumors. (23) Long-term prognosis for patients with intraventricular epidermoids and well-preserved neurological conditions is good, even in the case of subtotal excision. (9, 13)

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