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Case Report

Ectopic craniopharyngioma of the fourth ventricle in a patient with Gardner syndrome

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ABSTRACT

Ectopic craniopharyngioma is uncommon and a craniopharyngioma confined purely within the fourth ventricle is extremely rare. We report a craniopharyngioma of the fourth ventricle in a 20-year-old man with Gardner syndrome. Imaging characteristics of craniopharyngiomas and fourth ventricle lesions are discussed with a review of the literature regarding the pathogenesis of craniopharyngiomas and the possible association with Gardner syndrome.

1. Introduction

Craniopharyngiomas are slowly growing tumors [1], classified as grade I tumors by the World Health Organization (WHO). Craniopharyngiomas commonly occur in the suprasellar region [2] because they arise from epithelial remnants anywhere along the obliterated craniopharyngeal tract from Rathkés cleft to the floor of the third ventricle [3].

Ectopic craniopharyngiomas have been reported in different locations, including temporal lobe [4], clivus, and posterior cranial fossa [5–7]. Ectopic craniopharyngiomas in the fourth ventricle are exceedingly rare and there are only four case reports of this occurrence [5,8–10]. There are several case reports of patients with Gardner syndrome who had craniopharyngiomas in the cerebellopontine angle [11–14]. To our knowledge, this is this first case of an ectopic fourth ventricle craniopharyngiomas in a patient with Gardner syndrome.

2. Case report

A 20-year-old man with Gardner syndrome, an autosomal dominant disorder caused by mutation in the adenomatous polyposis coli gene APC on chromosome 5q, characterized by multiple polyposis predisposing to carcinoma of the colon and craniomaxillofacial osteomas, epidermoid cysts, and fibromas, presented with a 1-month history of headache. The patient had history of hepatoblastoma in infancy that was treated with partial right hepatectomy and subsequently followed by surveillance computed tomographic imaging (CT) and ultrasound of the abdomen, with no evidence of recurrence. His past surgical history also included colectomy at 15 years of age, which was done as a preventative measure because, by age 35 years, 95% of individuals with familial adenomatous polyposis (FAP) develop colon cancer [15]. At presentation, the patient was not taking any medications. The patient denied smoking tobacco, consuming alcohol, or recreational drug abuse. Physical examination at presentation revealed normal vital signs. He was alert and oriented to time, place, and person. Speech was clear and language was intact. On the neurological examination, cranial nerves II–XII were intact. Strength was full in all extremities and tone was normal. The laboratory values including complete metabolic panel, complete blood count and urinalysis, and cultures were within normal limits. Per patient's history, he had CT of the head at an outside institution, where a soft tissue mass in the fourth ventricle was reported; however, images were not available to us for review. Brain magnetic resonance imaging (MRI) with contrast was performed and revealed a mass in the fourth ventricle with irregular borders measuring 1.7 cm in maximum diameter. On the precontrast T1-weighted images

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(T1WI), the mass was predominantly T1 hypointense with respect to brain parenchyma with several rounded foci of T1 shortening characterized by bright signal (Fig. 1A). Postcontrast T1WI showed nodular enhancement of the solid components of the lesion (Fig. 1B). On T2-weighted images (T2WI), the mass was hyperintense relative to brain parenchyma except for several rounded foci of signal hypointensity (Fig. 1C). Gradient echo (GRE) sequence showed foci of susceptibility that were suggestive of calcification versus hemorrhage (Fig. 1D). Diffusion-weighted images (DWI) did not show abnormal diffusion restriction. There was no hydrocephalus. The patient also underwent MRI of the cervical, thoracic, and lumbar spine that was within normal limits.

The patient underwent tumor resection via a suboccipital craniotomy. Intraoperative stereotactic localization was used to determine the best surgical approach. It was determined that access through the vermis was the best route to the tumor. A midline incision in the middle third of the dorsal vermis was made and carried down to the white matter. Care was taken to leave the inferior portion of the vermis intact. The tumor was gray-green in color and was mildly to moderately vascular. The tumor was adherent to the inferior medullary velum and was not adherent to the brain stem, the floor of the fourth ventricle, or foramen of Luschka. The tumor was then gently removed from the cavity of the fourth ventricle. There was a small amount of bleeding associated with the base of the tumor in the region of inferior medullary velum that was easily controlled with bipolar electrocautery. After resection of the tumor, surgical cavity was meticulously inspected for hemorrhage and residual tumor; neither was identified.

The patient tolerated the procedure well and was discharged to subacute rehabilitation facility a few days postprocedure. Immediate postoperative MRI and the subsequent MRI performed at 3 months revealed no evidence of residual or recurrent tumor.

Histopathologic evaluation of the tumor (Fig. 2) was diagnostic of adamantinomatous craniopharyngioma and revealed a multicystic epithelial neoplasm with sheets and trabeculae consisting of basally palisading cells separated by regions of spindled and epithelioid squamous cells. Nuclei were generally round or ovoid with rare isolated mitotic figures. Degenerating and ghosted squamous cells with nodular keratin and lamellar keratin deposits were present. There were clusters of macrophages and focal calcification. The adjacent neural parenchyma

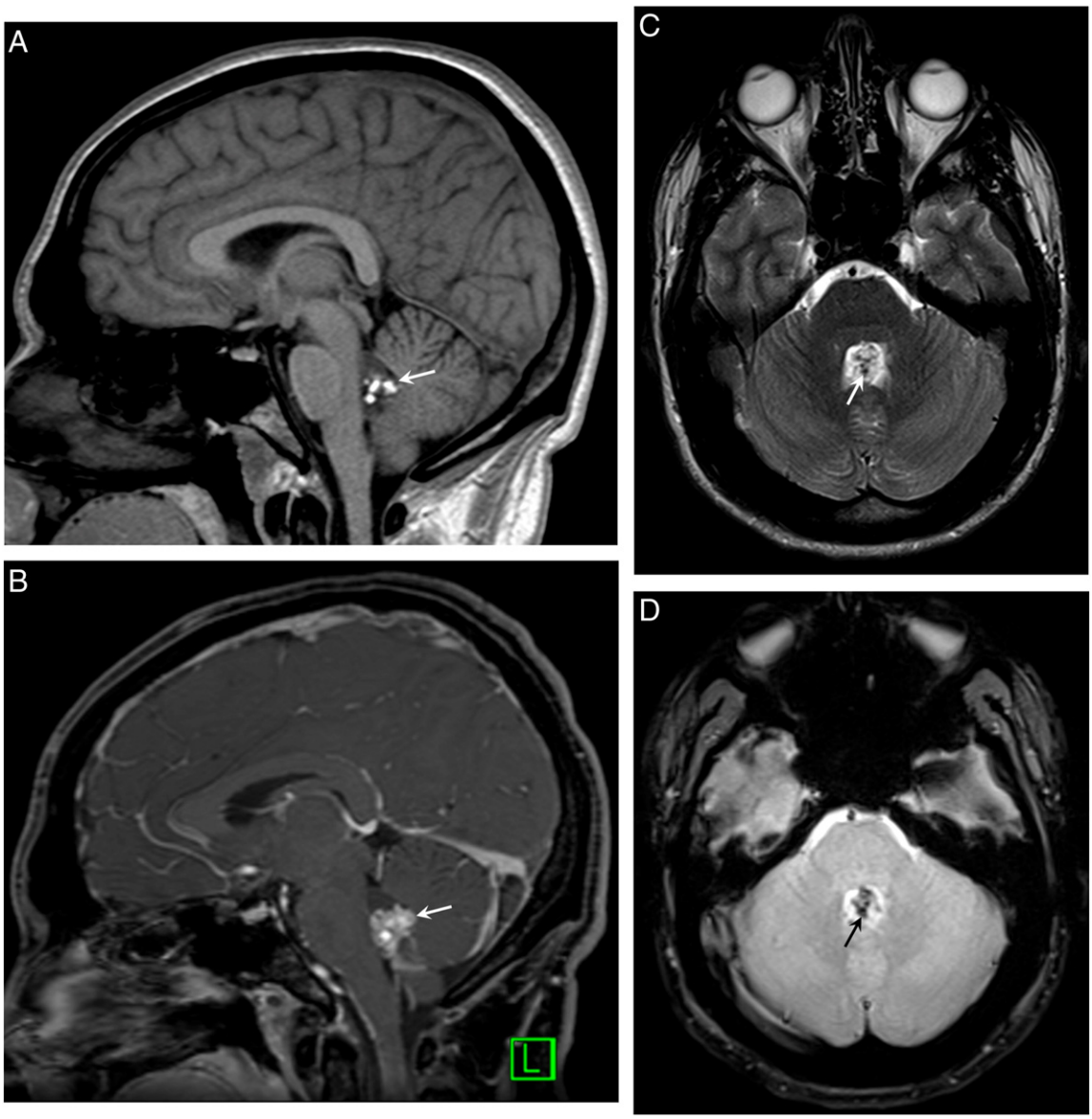

Fig. 1. MRI of the brain with contrast. (A) Sagittal precontrast T1WI, (B) sagittal postcontrast T1WI, (C) axial T2WI, and (D) axial GRE image show a mass in the fourth ventricle that is predominantly T1 hypointense to brain tissue with several foci of bright signal and nodular enhancement. The mass is T2 hyperintense with several hypointense foci of susceptibility that are more evident on GRE.