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Case Report

Ectopic Choroid Plexus Papilloma

Abstract

Choroid plexus papilloma is a rare intracranial neoplasm. These lesions most commonly present in the fourth ventricle in adults and lateral ventricle in children. Herein, we report a case of a 57-year-old male with complaint of backache of few weeks duration. Magnetic resonance imaging revealed an intradural and extramedullary space occupying lesion at D8 level. Histology showed multiple free-floating papillae with a central fibrovascular core. The cells showed immunoreactivity for vimentin, glial fibrillary acidic protein, synaptophysin, S-100 protein, and cytokeratins (CK, CK7).

Keywords: *Choroid plexus papilloma, ectopic neoplasm, extramedullary, intradural*

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Introduction

Choroid plexus papillomas (CPPs) are benign and slow growing tumors (WHO Grade I).[1] Choroid plexus tumors comprise of 0.3–0.6% of all brain tumors. Of these, 2–4% account for those that occur in children under 15 years, and 10–20% of those manifesting in the 1st year of life.[2] These lesions frequently occur in the lateral ventricle in children and fourth ventricle in adults.[3] Very few cases have been reported of disseminated CPP. Dissemination is primarily within spinal subarachnoid space.[4] The overall male:female ratio is 1.2:1; for lateral ventricle tumors, this ratio is 1:1 and for fourth ventricle tumors, it is 3:2.[2] Total surgical resection is the treatment to avoid chances of recurrence. Herein, we report a rare case of intradural, extramedullary ectopic CPP at D8 level. To the best of our knowledge, only one case of CPP in the sacrospinal region has been reported in literature so far.[4]

Case Report

A 57-year-old male presented with backache of a few weeks duration. There were no other complaints. Magnetic resonance imaging (MRI) of dorsal spine with whole spine screening (plain and contrast) was performed, which showed a well-defined enhancing mass lesion 9 mm in diameter in the left posterior epidural space at the D8 level [Figure 1]. The dorsal cord was compressed and displaced anteriorly and to the right by the mass lesion. On the T1- and T2-W images, the lesion had mixed signal intensity. No other lesion was detected by MRI and positron emission tomography (PET) scan.

On histopathology, sections showed bands of fibrocollagenous tissue and a tumor composed of papillae lined by cuboidal to columnar epithelium with a central fibrovascular core with mild nuclear pleomorphism [Figure 2]. There was no evidence of necrosis or mitoses.

The tumor cells expressed vimentin, cytokeratins (CK), glial fibrillary acidic protein (GFAP), S-100 protein, synaptophysin and CK-7 (occasional) and were immunonegative for Epithelial membrane antigen (EMA), CK-20, thyroid transcription factor-1 (TTF-1), carcinoembryonic antigen (CEA), chromogranin, and Ber EP4. The MIB-1 labeling index was approximately 7–10% [Figures 3 and 4].

Based on these histological features and immunohistochemical profile, a diagnosis of CPP (WHO Grade I) was given.

The tumor was completely excised and the patient is following up for 2 years with no recurrence or tumor at any other site.

Discussion

CPP is a histologically benign lesion included in WHO Grade I. It is known to arise primarily in the lateral or fourth ventricle which is the normal location of choroid plexus. There are many cases of CPP documented in the literature,

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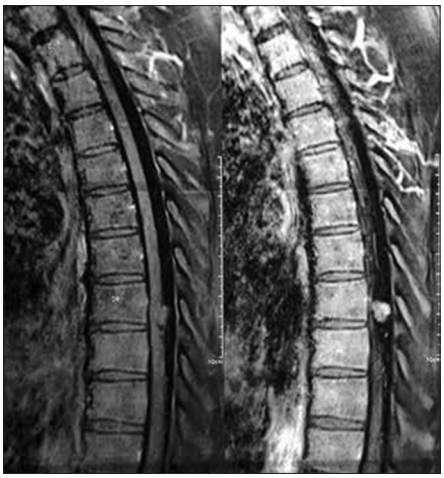

Figure 1: Magnetic resonance imaging showing a well-defined, enhancing lesion 9 mm in diameter at left posterior epidural space at D8 location displacing and compressing the cord

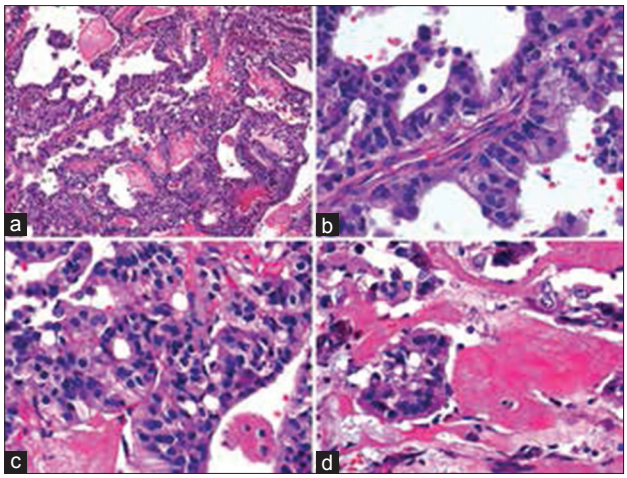

Figure 2: Multiple sections showing tumor comprising of papillae lined by cuboidal to columnar epithelium with a central fi brovascular core. Mild nuclear pleomorphism can be appreciated. Focal areas of fibrin seen. (a) H and E, ×100; (b-d) H and E, ×400

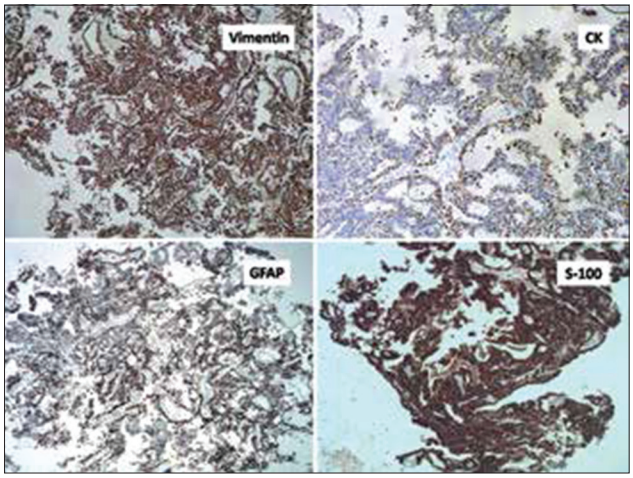

Figure 3: Tumor cells showing positivity for vimentin, cytokeratins, fibrillary acidic protein and S-100 protein immunohistochemical stains

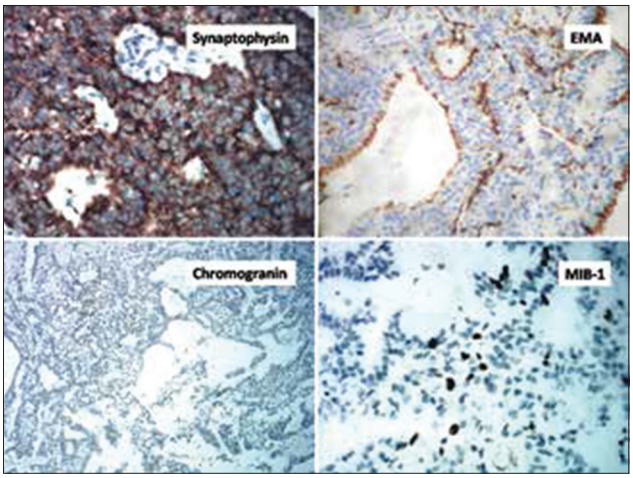

Figure 4: Tumor cells show positivity for synaptophysin, negativity for epithelial membrane antigen and chromogranin immunohistochemical stains. MIB-1 labeling index approximately 7–10%

some of which are at unusual sites [Tables 1 and 2]. Extra-ventricular occurrence of CPP is very rare.[5,6] Cerebellopontine angle is one of the unusual sites of CPP, which might occur due to direct tumor extension through fourth ventricle or foramen of Lushka.[3] Although benign, CPPs have been noted to disseminate via CSF and these, involve the subarachnoid space. The occurrence of CPP at an ectopic location like sacral canal has also been noted. There is a possibility that presence of such ectopic choroid plexus might be due to metaplasia of ependymal rests.[4]

Most common presentation of CPP is hydrocephalus caused by increased CSF production or obstruction to the CSF pathway. These are benign lesions, but their dissemination at the distant site has been noted.[7] This dissemination is mainly through CSF and involves subarachnoid space.

On noncontrast head computed tomography, CPPs are isodense to hyperdense as compared with brain parenchyma. Internal calcification is present in up to 20% of cases. On MRI, CPPs are typically isointense to hyperintense on T2 and isointense to hypointense on T1.[6]

Macroscopically, CPPs are cauliflower-like masses which are well-circumscribed. It may be adherent to the ventricular wall but usually well-delineated from the ventricular tissue.[2] Microscopically features are similar to normal choroid plexus having fibrovascular connective tissue lined by single layer of cuboidal to columnar epithelial cells having round to oval basal nuclei. Cells tend to be more crowded, elongated or stratified. There is no conspicuous mitotic activity or necrosis.

On immunohistochemistry, CPPs are positive for CK, CK7, synaptophysin, vimentin, S-100 protein and GFAP, all of which are consistent with our case. S-100 protein is present in 55–90% of cases. There is a report stating staining for synaptophysin to be strongly positive in both normal and neoplastic choroid plexus epithelial cells, which is not

Table 1: Distribution of choroid plexus papilloma at unusual sites

Cases	Age group (years)	Sex	Unusual sites
Cai *et al*., 2015	7	Female	Cerebellar vermis
Sasani *et al*., 2014	9	Female	Pineal region
Xiao *et al*., 2013	10	Male	Brainstem (pons)
Bian *et al*., 2011	31	Female	Pituitary fossa
Kinoshita *et al*., 2010	52	Female	right cerebellomedullary cistern
Sameshima *et al*., 2010	51	Female	Sella turcica
Tuchman *et al*., 2009			Cavum septum pellucidum
Boldorini *et al*., 2009	60	Female	Sacral nerve roots
Ma *et al*., 2008	49	Female	Sella turcica
Kurtkaya-Yapicier *et al*., 2002	50	Female	Sacral canal

[Cai *et al* - Clin Neuropathol. 2015 May-Jun;34(3):132-5, Sasani *et al* - Childs Nerv Syst. 2014 Jul;30(7):1307-11, Xiao *et al* - J Neurosurg Pediatr. 2013 Sep;12(3):247-50, Bian *et al* - Acta Neurochir (Wien). 2011 Apr;153(4):851-7 , Kinoshita *et al* - Neurol Med Chir (Tokyo). 2010;50(10):930-3, Sameshima *et al* - Neurol Med Chir (Tokyo). 2010;50(2):144-6, Tuchman *et al* - J Neurosurg Pediatr. 2009 Dec;4(6):580-3, Boldorini *et al* - J Neurosurg Spine. 2009 Jan;10(1):51-3.Ma *et al* - J Neurooncol. 2008 May;88(1):51-5]

Table 2: List of reported choroid plexus papilloma over the last 10 years

Year	Age group	Sites
2015	5-50 years	Lateral ventricle, third ventricle, CP angle, extra-ventricular and intra-parenchymal, cerebellar vermis
2014	3 weeks-52 years	Posterior third ventricle, lateral ventricle, CP angle, pineal region
2013	7 weeks-41 years	Third ventricle, CP angle, brainstem (pons), lateral ventricle, fourth ventricle
2012	6-72 years	Fourth ventricle, lateral ventricle
2011	26-42 years	Cerebral parenchyma, fourth ventricle, pituitary fossa
2010	5 months-59 years	CP angle, cerebellomedullary cistern, fourth ventricle, sella turcica
2009	7 months-62 years	Cavum septum pellucidum, lateral ventricle, sacral nerve roots, fourth ventricle
2008	38 weeks-11 years	Sella turcica, lateral ventricle, CP angle
2007	2 months-51 years	Posterior fossa, fourth ventricle, third ventricle, lateral ventricle
2006	1-49 years	Third ventricle, posterior fossa
2005	54 years	Posterior fossa

CP – Cerebellopontine

confirmed by others.[2] Prominent staining for EMA is typically not found.

The treatment of choice of CPP is total surgical excision of the tumor. In young children, complete resection is sometimes difficult because of the typically large size of the tumor and vascularity.[8] Complete removal of tumor results in long-term survival rates and adjuvant treatment is not required.[9]

The case presented here demonstrates the unusual occurrence of CPP in extramedullary and intradural location at D8 level. Although the site is extremely rare, the diagnosis is established on the basis of histological and immunohistochemical features.

Taking patient's age into consideration, metastatic papillary carcinoma may be considered as one of the differential diagnosis. However, this can be efficiently ruled out in view of characteristic histological features, immunopositivity for vimentin, CK, GFAP, S-100 protein, synaptophysin, and immunonegativity for EMA, CK-20, TTF-1, CEA, and Ber EP4. Some studies have shown evidence of strong positivity for synaptophysin in choroid plexus lesions, which helps to differentiate it from metastatic papillary carcinomas.[10] Complete PET scan was performed, which did not show evidence of any other primary lesion, again favoring it to be a nonmetastatic lesion.

As this is a solitary lesion, this case is less likely to be a disseminated malignancy.

In summary, we report a rare case of intradural, extramedullary CPP at D8 level. Looking at the classic histology of the lesion, its extracranial location and absence of neoplastic process at any other site, this is most likely to be a case of ectopic CPP.

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Conflicts of interest

There are no conflicts of interest.

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