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Choroid Plexus Papilloma Presenting at Cerebellopontine Angle – A Case Report

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Abstract **Case Report**

Choroid plexus papilloma is slow growing benign tumor of neuroepithelial origin. This tumor accounts for 0.4% to 1% of intracranial neoplasm. These tumors are more common in children with incidence of 1.5% to 4% and rare in adults. Lateral ventricles are common sites in children and 4th ventricle is common site in adults. Cerebellopontine angle is rare site of occurrence of this tumor. These tumors have tendency to recur after surgical resection and also to metastasize, though they are benign. We report a case of choroid plexus papilloma presenting at cerebelloportine angle in a 45-years male.

Keywords: Choroid plexus papilloma, cerebellopontine angle, intraventricular tumor.

INTRODUCTION

Choroid plexus papilloma is a rare slow growing benign intracranial tumor arising from the lining epithelium of choroid plexus. They comprise 0.4% to 1% of intracranial neoplasms. These tumors occur more commonly in children than in adults [1]. As these tumors arise from choroid plexus tissue they are commonly found in lateral ventricles in children and fourth ventricles in adults. Third ventricle and cerebellopontine angle are rare sites for choroid plexus papillomas. [2] Though the tumors are benign in behaviour they have the tendency to metastasize. These tumors can also undergo malignant transformation. We report a case of choroid plexus papilloma at the cerebellopontine angle in a 45 years male patient presenting with features of brain stem compression.

CASE REPORT

A 45 years male patient presented to neurosurgery department with chief complaint of decreased bilateral visual acuity. Computerised tomography of the brain showed right cerebellopontine angle tumor presenting as irregular heterogeneous predominantly hyper dense areas measuring 4x2.5cm in the right cerebellopontine angle involving vermis with perilesional edema and mass effect in the form of effacement of 4th ventricle. 3rd ventricle appears prominent.

Magnetic resonance imaging brain showed cerebellopontine angle intracranial space occupying lesion with a well-defined heterogeneous T1 iso to grey matter. T2 hyper intense lesion not showing diffusion restriction with increased apparent diffusion coefficient (ADC) signal and few areas of blooming on Susceptibility weighted angiography (SWAN) measuring 3.5x2cm with mild peripheral edema noted in right cerebellopontine angle. Mass effect is noted in the form of compression of pons, medulla, cerebellar hemisphere and fourth ventricle on right side. Clinical diagnosis of meningioma or schwannoma was made.

Patient underwent right sided suboccipital clavectomy by rectosigmoid approach. Friable and vascular lesion was present at the right angle. Tumor was removed and sent for histopathological examination. Grossly we received multiple grey white soft tissue bits altogether measuring 5X4cms. Microscopic examination revealed lesion composed of papillae with delicate fibrovascular core. Papillae were lined by cuboidal and low columnar epithelium (Figure1,2).

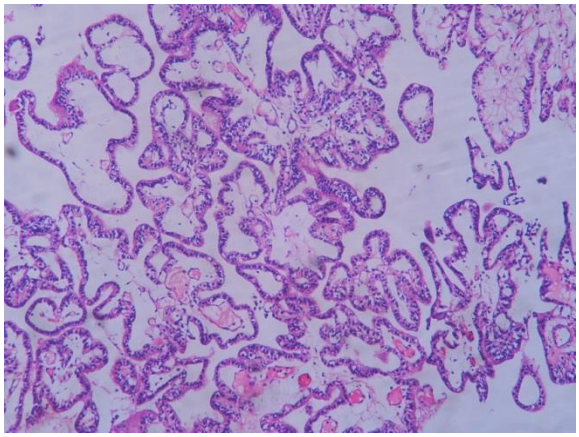

Figure 1: Lesion composed of papillae having delicate fibrovascular core lined by low columnar lining epithelium (H&E, X100)

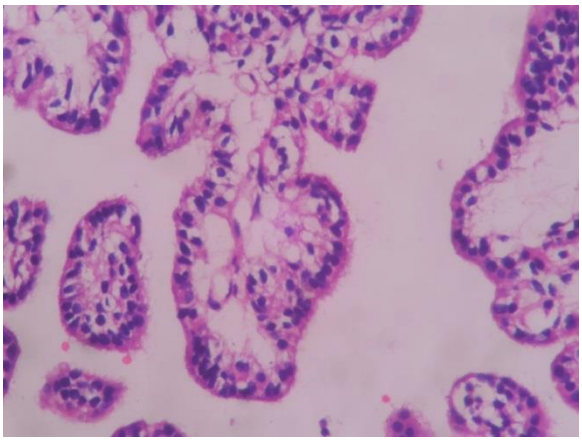

Fig-2: Lesion composed of papillae having delicate fibrovascular core lined by cuboidal to low columnar epithelium having a linear face (H&E, X400)

DISCUSSION

Choroid plexus papilloma is a rare benign slow growing intracranial tumor comprising 0.4% to 1% of all intracranial neoplasms. They arise from cuboidal epithelium lining the choroid plexus. Choroid Plexus Papillomas are more common in the lateral ventricles in children and in 4th ventricles in adults. Rarely tumor arises at the cerebellopontine angle as a direct extension of this neoplasm at the foramen of Luschka or from the metastatic seedling through cerebrospinal fluid pathway.

Common clinical presentation of the cerebellopontine angle lesions are dysfunction of hearing and facial movement, dizziness, headache, ataxia, hydrocephalus and papilledema. Our case presented with decreased bilateral visual activity.

Lesions which simulate Choroid plexus papilloma arising at cerebellopontine angle in their clinical presentation are vestibular schwannoma, meningiomas and epidermoid cyst. However imaging studies may primary help in providing preopoeratic diagnosis [3]. On non-contrast Computerized Tomography (CT) Choroid plexus papilloma are hyperdense to isodense when compared with brain parenchyma. 20% of cases show internal calcification [4]. The CT picture of choroid plexus papilloma occurring in the intraventricular sites is homogenously enhancing lobulated mass having cauliflower like appearance showing frond like irregular pattern.

On MRI these tumors are isotense to hypertense on T2. On T1 Choroid plexus papilloma appear isotense to hypotense [5]. Calcification and infarction can occur in tumor. In our case heterogeneous T1 lesion was seen on MRI which is uncommon.

Grossly the tumors are lobular, well circumscribed and have broad compressive front. Microscopically, these tumors show papillae with delicate fibrovascular core. Papillae are lined by an orderly layer of columnar epithelium present on basement membrane which has a linear face when compared to “hobnail” or “cobblestone” appearance of normal tissue. Tumor cells can also form tubules or acini. Other changes which can occur in tumor are oncocytic change, Pigmentation, Xanthamatous change, calcification, cartilage or bone. Choroid plexus neoplasms having well differentiated epithelium but scattered mitosis (2 or more/10hpf) which are difficult

to categorize in a papilloma or carcinoma are labelled as atypical choroid plexus papilloma [6]. Choroid plexus carcinoma can be differentiated from choroid plexus papilloma by the features like blurring of the papillary features, cellular pleomorphism, increased atypical mitotic activity, areas of necrosis and invasion into brain parenchyma [7]. Immunohistochemically tumor cells are reactive for Vimentin, Cytokeratins, S-100 protein and Synaptophysin. Few cases may show reactivity for EMA and GFAP

Choroid plexus papilloma should be differentiated from papillary ependymoma and metastatic papillary carcinomas. In papillary ependymomas, epithelial appearance elements and non-epithelial glial cells with fibrillar background will be present where as highly fibrillated areas are not diagnostic of papillomas. Ependymomas lack prominent basement membrane and are strongly positive for GFAP.

Metastaic intraparenchymal papillary lesions are positive for antibodies BerEP4 & HEA 25.Metastatic pulmonary neoplasms are positive for Thyroid Transcription Factor-1(TTF-1) whereas Choroid plexus papilloma cells are negative.

The recommended treatment is complete microsurgical excision of the tumor. Recurrences of these tumors can be reduced by radiation therapy.

CONCLUSION

Imaging characteristics of Choroid plexus papilloma occurring at cerebellopontine angle are not similar to those occurring in the intraventricular sites. On imaging these tumors have heterogenous enhancement which can be present in other tumors like Schwannoma and meningioma occurring at cerebellopontine angle. Choroid plexus papilloma should be considered as differential diagnosis for the tumors occurring at cerebellopontine angle.

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