

Primary Choroid Plexus Papilloma of the Cauda Equina. A Case Report

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Tumors arising from the choroid plexus are not common. They most often affect the pediatric population and account for less than 1% of all central nervous system tumors. While intermediate forms do exist, choroid plexus tumors are typically described as either papilloma or carcinoma, with the latter being encountered much less frequently. Their location is classically confined to regions where normal choroid plexus resides. We report a rare case of an adult female with a primary choroid plexus papilloma arising from her cauda equina.

CASE REPORT

History and Presentation. A 52-year-old female presented to neurosurgical attention with an 18-month history of progressive buttock pain radiating into her right leg. The pain would occasionally wake her from sleep and was aggravated by standing and walking. She did not complain of any weakness or numbness of her lower extremities, nor did she describe any bowel or bladder dysfunction. Her medical history consisted only of a benign thyroid lesion which was surgically resected 11 years prior to her presentation.

Examination. Her physical examination was unremarkable. She did not have motor or sensory deficits and her deep tendon reflexes were present and symmetrical. Magnetic resonance imaging (MRI) investigation of her lumbar spine revealed a well-defined, 2.5 cm intradural lesion at the L4-L5 level (Figure 1). The lesion was isointense on T1 imaging, mildly hyperintense on T2 imaging and showed homogeneous enhancement with gadolinium administration. The differential diagnosis consisted of meningioma, neurofibroma, schwannoma and ependymoma.

Operation. The patient underwent a complete L4-L5 laminectomy followed by a midline durotomy. An encapsulated, firm, multilobulated mass was encountered within the thecal sac. The tumor was adherent to the dura as well as the L4 and L5 nerve roots on the right. Portions of the tumor coursed along the nerve root into the neural foramen. At the superior aspect of the tumor it appeared to be arising from the filum terminale. The nerve roots were dissected off of the tumor capsule. The size of the tumor prevented en bloc resection. Internal debulking was performed initially to aid with dissection from the nerve roots. A gross total resection was achieved. There were no changes in the intraoperative neurophysiological monitoring throughout the case. Quick section diagnosis was consistent with ependymoma.

Pathological Findings. Microscopic examination revealed a tumor with a prominent papillary pattern (Figure 2). The papillary fronds were cuboidal-to-columnar cells which lined a fibrovascular stroma. Lining of tumor cells against a neuropil was not demonstrated. The cells were predominantly

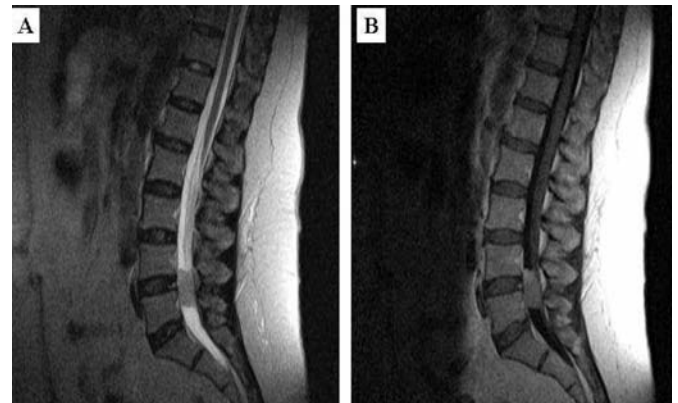


Figure 1: Mid-sagittal MRI investigation of the thoracic and lumbar spine. T2-weighted imaging reveals an intradural tumor at the L4-L5 level (A). The tumor shows homogeneous enhancement on T1-weighted imaging with gadolinium administration (B).

monomorphic occasionally containing vacuoles. Extensive immunohistochemical study revealed strong reactivity for CK7, CK20, low molecular weight keratin, S100 protein, GFAP, MAP2, BCL-2, CD56, TAB250 and SP3. Focal positivity of EMA was seen along the borders of the papillae. Though somewhat variable, CD99 and NSE, were also reactive. Nestin studies showed both a reactive and non-reactive population of cells within the tumor. Tumor cells were non-reactive for TTF-1, thyroglobulin and CEA. Mitotic figures were exceedingly rare and the MIB-1 index was less than 2% (Figure 3). Given the phenotypic and immunohistochemical profile of this tumor an initial diagnosis of low-grade papillary spinal tumor was made. Notwithstanding the location of the tumor the diagnosis of choroid plexus papilloma was favored.

Postoperative Course. The patient's pain improved after surgery. She continued to have a normal neurological examination, including her bowel and bladder function. Her

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hospital stay was uneventful and she was discharged home on the third postoperative day. Given the diagnosis of choroid plexus papilloma a contrast enhanced MRI of the brain was performed to rule out the possibility of a drop metastasis from an intracranial choroid plexus lesion. No intracranial pathology was identified. She was referred to the oncology service for an opinion regarding management of this interesting tumor. They opted to observe the patient with serial clinical and radiological examinations. The patient has undergone contrast enhanced MRI investigations of the lumbar spine every three to six months for approximately three years. Early postoperative studies revealed mild nodular enhancement of the cauda equina and questionable new enhancement of the cord at T11-12. These findings have been stable on serial imaging.

DISCUSSION

Choroid plexus tumors are rare. They account for 0.3-0.6% of all brain tumors¹. Their average annual incidence is approximately 0.3 per million people and they occur most commonly in childhood, particularly in the first decade of life¹⁻⁵. The World Health Organization now recognizes three types of choroid plexus tumors – papilloma (CPP), atypical papilloma and carcinoma (CPC), with CPP outnumbering CPC by a ratio of 5:1^{1,2}.

Choroid plexus tumors are typically confined to the regions of the central nervous system where choroid plexus is found. Approximately half of all choroid plexus tumors are located in the lateral ventricles, with another 40% in the fourth ventricle and 5% in the third ventricle^{1,3,6}. Multiple ventricles are involved in 5% of cases^{1,3,6}. Given their predilection to the ventricular system cerebrospinal fluid (CSF) seeding and so-called "drop metastasis" are occasionally encountered⁷⁻⁹. Less frequently seen are cases of de novo intracranial extraventricular choroid plexus tumors of the cerebellopontine angle (CPA)¹⁰⁻¹², foramen magnum¹³, suprasellar region¹⁴, or parenchyma¹⁴. Nearly all reported cases of choroid plexus tumors involving the spinal canal involved patients with a known intraventricular tumor⁷⁻⁹.

A primary choroid plexus papilloma of the spinal canal, as described here, is exceedingly rare. Our literature review identified only two similar cases^{15,16}. Kurtkaya-Yapici et al were the first to report on an extradural choroid plexus papilloma of the sacral canal in a 50-year-old woman¹⁵. She underwent gross total resection of the tumor and there was no evidence of radiographic recurrence after nine months of follow-up. Boldorini et al. recently presented a case of a 60-year-old woman with a primary choroid plexus papilloma of the sacral nerve roots¹⁶. Their patient also underwent gross total resection with no evidence of recurrence at one year postoperatively.

The microscopic appearance of the tumor in our case led us to revise our initial differential diagnosis. Given the prominent papillary pattern the following potential pathologies were entertained – primary choroid plexus papilloma, myxopapillary ependymoma, papillary ependymoma, metastatic papillary carcinoma, papillary glioneuronal tumor, papillary tumor of the pineal gland, choroid plexus papilloma secondary to drop metastasis. The latter three were definitively ruled out by virtue of the normal MRI investigation of the brain. Given the cellular monomorphism, paucity of mitotic figures, low proliferative index and negative immunostaining for thyroglobulin, TTF-1

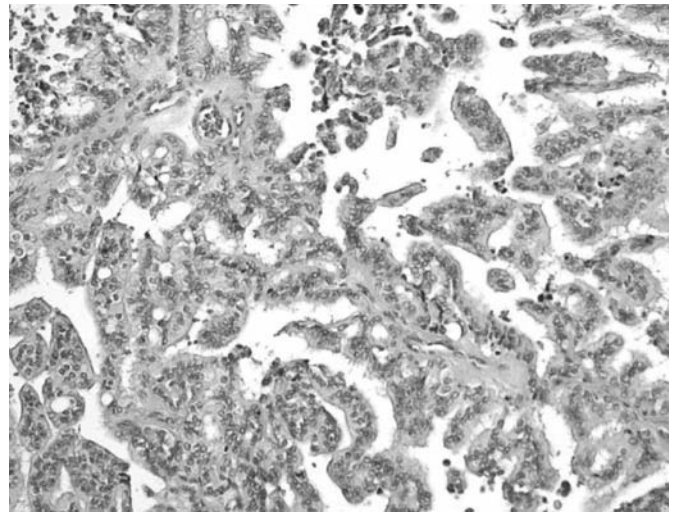


Figure 2: Microscopic images of the tumor reveal a prominent papillary pattern (hematoxylin and eosin; 10X). Cuboidal-to-columnar cells are seen lining a fibrovascular stroma.

and CEA the possibility of metastatic disease could also be excluded. Furthermore, the high expression of cytokeratins and EMA in carcinomas is rarely associated with immunostaining for other proteins such as S-100 protein, GFAP and NSE as described in the present case. While myxopapillary ependymomas are also characterized by tumor cells radially arranged around a stromal core, the absence of myxoid matrix material and strong cytokeratin immunopositivity excludes this diagnosis as well¹⁷. Papillary ependymomas are rare tumors which also share a similar microscopic appearance to the tumor presented here. Contrary to our case however, their stromal core consists of glial cells (versus connective tissue) and lacks a basement membrane. Furthermore, they are non-reactive for synaptophysin. Notwithstanding the location of the tumor presented here, the morphological and immunohistochemical characteristics are consistent with a primary choroid plexus papilloma of the cauda equina.

Hypotheses in the literature to explain the origin of a primary choroid plexus papilloma within the spinal canal are scarce given the novelty of the diagnosis. The notion of ectopic choroid plexus has been described to explain the presence of choroid plexus tumors with no connection to the ventricular system^{14,15}. Choroid plexus tissue has been described in cysts of the CPA, oropharynx and supratentorial cerebrum¹⁸⁻²⁰. The other hypothesis set forth by Kurtkaya-Yapici et al. was that of choroid plexus metaplasia of ependymal cells¹⁵. Our tumor specimens did not contain any normal ependyma or choroid plexus tissue, making each of these hypotheses unlikely.

In conclusion, this case illustrates a low-grade papillary neoplasm of the cauda equina whose characteristics are consistent with a choroid plexus papilloma. While the location of such a tumor is extremely rare there is evidence in the literature to support it. Existing hypotheses to account for the "ectopic" nature of such a tumor unfortunately cannot adequately explain the case at hand.

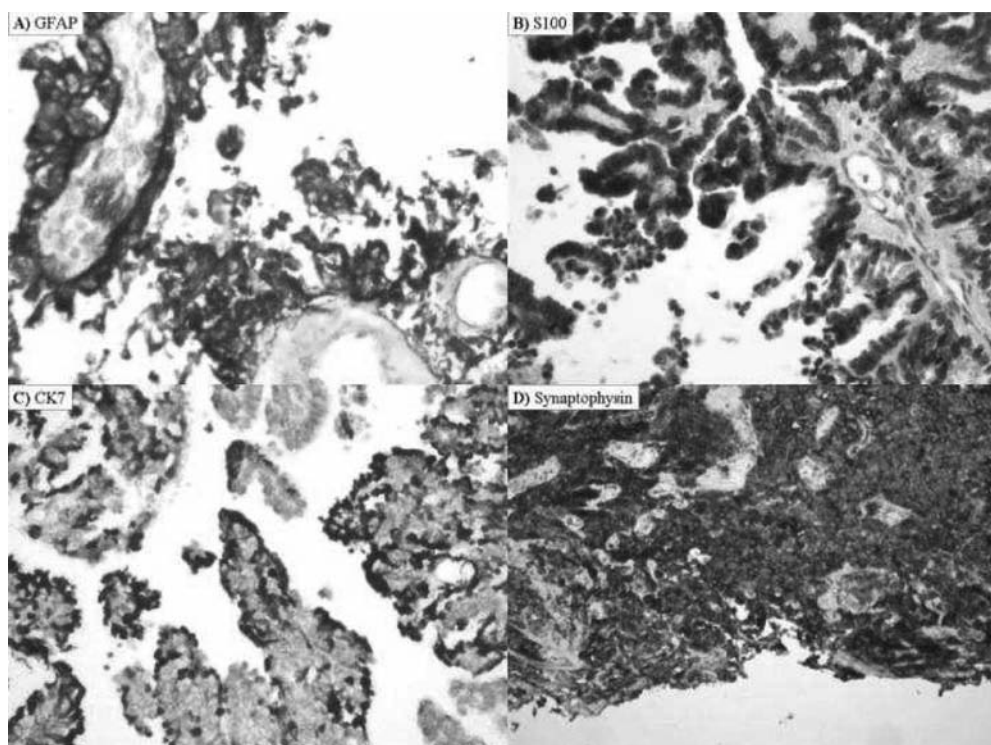


Figure 3: Immunohistochemical analysis demonstrates a staining pattern consistent with a choroid plexus papilloma. GFAP (A; 40X), S100 (B; 20X), CK7 (C; 20X) and synaptophysin (D;10X) all show strong positivity.

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