Delayed occurrence of multiple spinal drop metastases from a posterior fossa choroid plexus papilloma

Case report

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✓ Choroid plexus papilloma is a benign central nervous system tumor that occasionally spreads along the subarachnoid space. The authors report the case of a 49-year-old man who presented with back pain 19 years after resection of a posterior fossa choroid plexus papilloma. Magnetic resonance imaging revealed multiple spinal lesions without any residual or recurrent intracranial tumor. All spinal lesions were resected and histologically diagnosed as atypical choroid plexus papilloma. The authors suggest that patients in whom choroid plexus papilloma is diagnosed should undergo total neuraxis imaging at the time of initial diagnosis as well as periodic follow-up examinations after resection to rule out drop metastases.

KEY WORDS • choroid plexus papilloma • central nervous system tumor • spinal metastasis

CHOROID plexus papillomas are rare benign tumors of the central nervous system that occur mostly in children and have a slight male predominance. They account for fewer than 1% of all intracranial tumors. These tumors are usually found in the left lateral ventricle in children and the fourth ventricle in adults. Other less common sites include the third ventricle and cerebellopontine angle. Papillomas may also occur extraventricularly as a result of direct extension or seeding within the intraventricular or subarachnoid space. We report a case of choroid plexus papilloma drop metastases in a man who presented almost 20 years after gross-total resection of his intracranial primary tumor.

Case Report

History. This 49-year-old man presented to our center with a 1-year history of sharp low-back pain, which radiated down both hips and extended to his right knee. He denied any bowel or bladder dysfunction. The patient had undergone gross-total resection of a posterior fossa choroid plexus papilloma 19 years previously at another institution. He had received neither chemotherapy nor radiation therapy following the resection of his intracranial tumor.

Examination. Motor, sensory, and reflex examination yielded normal findings, with the exception of numbness in the left fourth and fifth toes. Magnetic resonance imaging of his lumbar spine showed a large intradural enhancing mass extending from the conus medullaris to the terminal filum (Fig. 1 A). Subsequent MR imaging of the rest of his spine revealed additional extramedullary enhancing lesions at the levels of C3–4 and T-7 (Fig. 1B and C). No recurrent or residual tumor was visible on MR images of the brain (Fig. 1D).

Operation. Pathological examination of a biopsy specimen of the lumbar lesion revealed findings consistent with choroid plexus papilloma. The patient underwent staged resection of all three spinal lesions. The cervical and tho-

Abbreviation used in this paper: MR = magnetic resonance.
racic lesions were removed entirely; the lumbar lesion, however, was only debulked due to its adherence to the nerve roots. Histological examination of all three lesions showed a differentiated papillary neoplasm with moderate pleomorphism and no mitoses, necrosis, or invasion. The final diagnosis of atypical choroid plexus papilloma was based on the findings of mild cytological pleomorphism and a slight increase in cellularity (Fig. 2). No sections of the intracranial lesion resected 19 years previously were available for review.

Postoperative Course. The patient’s back and leg pain resolved; however, he experienced urinary retention, which was persistent at his most recent follow-up examination (2 months after surgery), as well as transient and mild lower-extremity weakness and transient saddle anesthesia.

Discussion

Choroid plexus papillomas arise from the epithelial cells of the choroid plexus lining the ventricles. Histologically, they consist of papillae formed by a single layer of columnar epithelial cells supported by a basement membrane overlying connective tissue. They are classified as World Health Organization Grade I tumors. Because of the benign classification of these tumors, total surgical resection is typically thought to be curative. Recurrence has been reported, however, even after gross-total resection, and when choroid plexus papilloma recurs, the tumor can undergo malignant transformation. The malignant form, choroid plexus carcinoma, is classified as a World Health Organization Grade III tumor and is characterized by anaplasia, mitosis, nuclear pleomorphism, necrosis, and invasion. Adjuvant radiation therapy and chemotherapy are recommended for patients with choroid plexus carcinoma but not for those with choroid plexus papilloma. The histological characteristics of the tumors have prognostic significance because the 10-year survival rates following surgical resection are 77% for choroid plexus papilloma and 35% for choroid plexus carcinoma. The term “atypical choroid plexus papilloma” is not well defined because of the paucity of reported cases. The diagnosis is used to indicate papilloma with only one or two malignant features, and it is associated with a prognosis that lies somewhere in the middle of the prognostic spectrum of choroid plexus tumors.

Craniospinal spread is a common feature of choroid plexus carcinoma, not papilloma, but a few cases of drop metastases from benign papilloma have been reported. Such metastases appear to be particularly associated with primary tumors in the posterior fossa. Drop metastases are usually found within a few years of diagnosis of the primary intracranial tumor. To our knowledge, this is the first reported case of spinal metastases from a choroid plexus papilloma detected as long as 19 years after the resection of the original intracranial tumor. Furthermore, our patient did not experience a local recurrence of the primary intracranial lesion. Assuming that the patient’s spinal lesions resulted from subarachnoid space seeding from the primary intracranial tumor, the possibility does exist that they were present at the time that the intracranial tumor was detected. Unfortunately, when the patient initially presented with the intracranial lesion, MR images of his entire spine were not obtained. This case perhaps illustrates the need for obtaining spinal MR images to screen for metastases in patients with intracranial choroid plexus papilloma.

Conclusions

Choroid plexus papilloma is considered a benign tumor with a good long-term prognosis if total resection can be
achieved. Nevertheless, reports of local recurrence and craniospinal seeding suggest diversity in the natural history of this disease. The present report of drop metastases found so long after primary tumor resection and without primary tumor recurrence adds to our understanding of the complexity of this tumor’s behavior. Further elucidation of the biology of choroid plexus papilloma will lead to a clearer understanding of the natural history of this neoplasm. Occurrence of drop metastases, although rare, suggests the need for total neuraxis imaging at the time of diagnosis. For cases in which the original staging study is negative and a complete resection is achieved, however, the yield of serial postoperative imaging is unknown. A reasonable recommendation might be to obtain a follow-up imaging study a year or two after the initial diagnosis.

References


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