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Case Report

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Isolated Lumbar Atypical Choroid Plexus Papilloma: A Case Report

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ABSTRACT

AIM: To report the first case of an isolated lumbar grade II atypical choroid plexus papilloma (CPP).

CASE REPORT: A 42-year-old man was admitted to the hospital because of back and leg pain. No urinary or rectal dysfunction was detected. Lumbar magnetic resonance imaging (MRI) showed a well-circumscribed, contrast-enhancing, intradural extramedullary mass at L2-3. He underwent L2 and L3 partial laminectomies for tumor resection and complete resection was achieved without causing neurological deficit. Histopathologic examination of the tumor resulted in a diagnosis of grade II atypical CPP. The Ki-67 staining index was 7%. No lesion was detected on postoperative craniospinal MRI.

CONCLUSION: Isolated lumbar atypical CPP in the lumbar region has not been previously reported. In the presence of a single spinal lesion, the diagnosis of CPP should be considered. Unlike metastatic and synchronous tumors, the pathogenesis of isolated choroid plexus tumors within the spinal canal has not been explained.

KEYWORDS: Atypical choroid plexus papilloma, Intradural, Lumbar, Single

ABBREVIATIONS: CPP: Choroid plexus papilloma, **MRI:** Magnetic resonance imaging

INTRODUCTION

Choroid plexus papilloma (CPP) is a rare neuroectodermal tumor found in the central nervous system. It is usually located in the lateral ventricles in children and the fourth ventricle in adults. Approximately 25% of choroid plexus tumors are malignant; these tumors tend to recur locally and can spread within the cerebrospinal fluid space (4). CPP is rarely seen as a single lesion in the spinal region without being synchronous or metastatic. Spinal CPP can occur in three different ways: 1) as a direct extension of a primary intraventricular papilloma, 2) as a drop metastasis from an orthotopic tumor, and 3) by development from ectopic choroid plexus (1). We present a patient with an isolated lumbar grade II atypical CPP.

CASE REPORT

History

A 42-year-old man was admitted to the hospital because of a 1-month history of back and anterior leg pain. Urinary and rectal dysfunction was not detected.

Initial Imaging

Spinal magnetic resonance imaging (MRI) showed a regularly circumscribed enhancing lumbar lesion that was isointense on T1-weighted images and hypointense on T2-weighted images (Figures 1A-C, 2A, B).

Operation and Postoperative Imaging

The patient underwent partial laminectomy at L2 and L3 levels in the prone position. After opening the dura in a linear fashion, the lumbar spinal roots were seen to be displaced posterolaterally by the tumor, which was attached to a nerve

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root. The tumor was debulked under the microscope using an ultrasonic aspirator and bipolar electrocautery. After surgery, the patient had no neurological deficit. Postoperative craniospinal MRI showed a complete resection; no additional lesions were found (Figure 3A, B).

Pathological Findings

Histopathological examination provided a diagnosis of atypical CPP. The tumor was partially composed of papillary structures lined by cuboidal or columnar cells and partially demonstrated more solid areas with oncocytic cells (Figures 4,

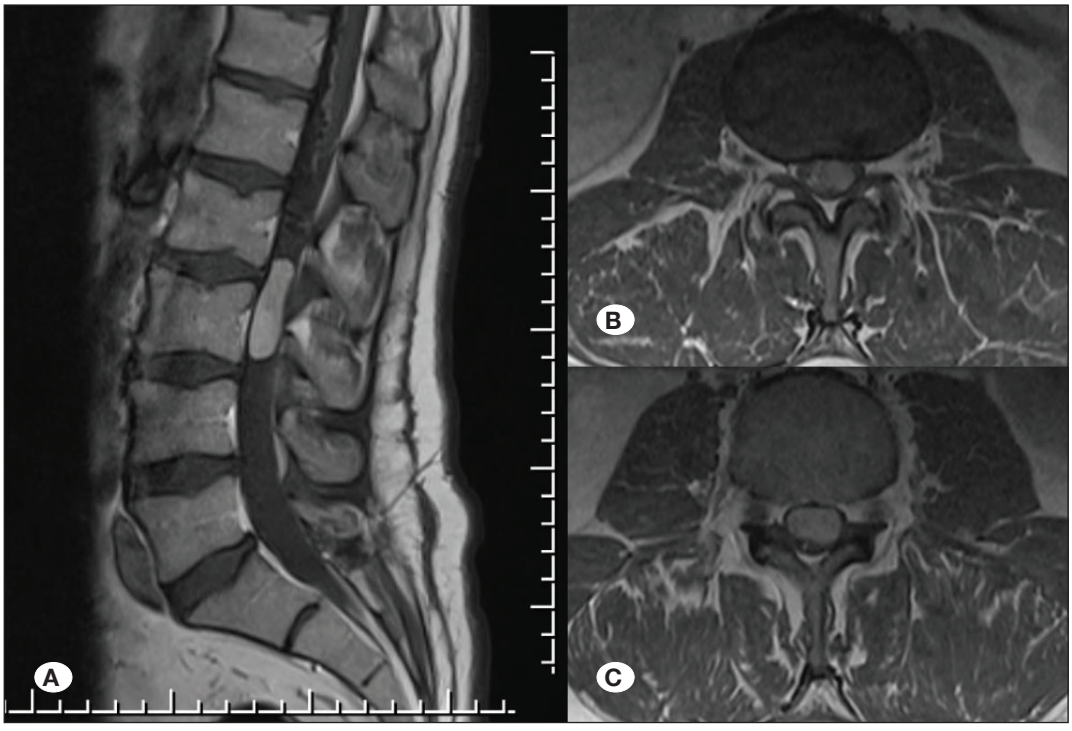

Figure 1: Contrast-enhanced lumbar magnetic resonance imaging shows a regularly circumscribed enhancing lesion **(A, B, C)**.

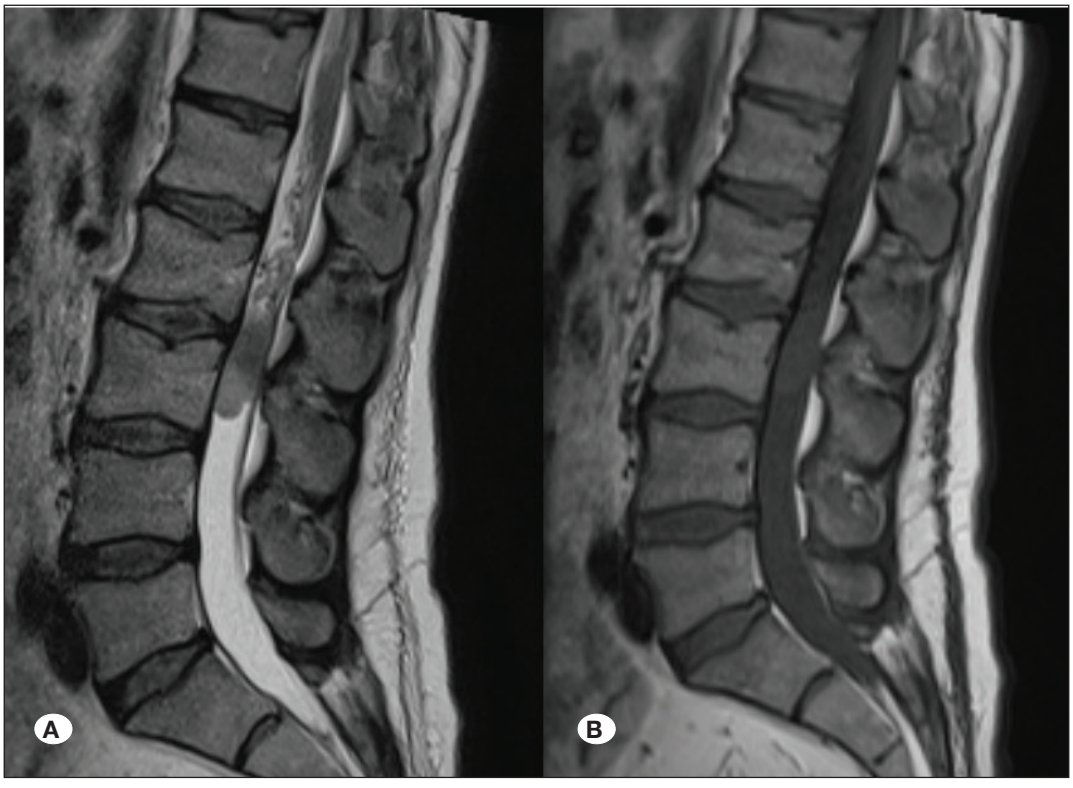

Figure 2: Lumbar magnetic resonance imaging shows the lesion was hypointense on T2-weighted sequences **(A)** and isointense on T1-weighted sequences **(B)**.

5). Mitoses were not detected. Immunohistochemical analysis demonstrated that tumor cells showed diffuse expression of cytoplasmic vimentin and synaptophysin (Figures 6, 7). Some tumor cells were positive for cytokeratin AE1/AE3 (Figure 8). Tumor cells were negative for glial fibrillary acidic protein, epithelial membrane antigen, S100, and carcinoembryonic antigen. MIB1 labeling index was 7%. Although the mitotic activity was lower than 2 mitoses per 10 high-power fields, the tumor was diagnosed as grade II atypical CPP based on the MIB1 labeling index.

DISCUSSION

CPP is rarely located in the spinal cord and cannot be radiologically distinguished from other spinal tumors (5). They are diagnosed after pathological evaluation. CPP is considered a grade I tumor with good prognosis and complete recovery after total excision is expected. Choroid plexus carcinoma is considered a grade III malignant tumor. In between the two is grade II atypical CPP, which may have the following features: increased mitotic activity, immunoreactivity for cytokeratin 7, and neurofilament protein-positive fibrils indicative of infiltration (4).

Kurtkaya-Yapıcıer et al. reported an extradural sacral CPP that was attributed to ectopic neuroectodermal residue and metaplasia of ependymal remnants that occurred during embryogenesis (7). Boldoroni et al. reported a CPP originating from sacral nerve roots. They also thought that the tumor might have originated from ependymal choroid plexus metaplasia (1). In our case, the tumor was intradural, had no primary connection with other ectodermal tissues in the sacral region, and had grade II histopathology.

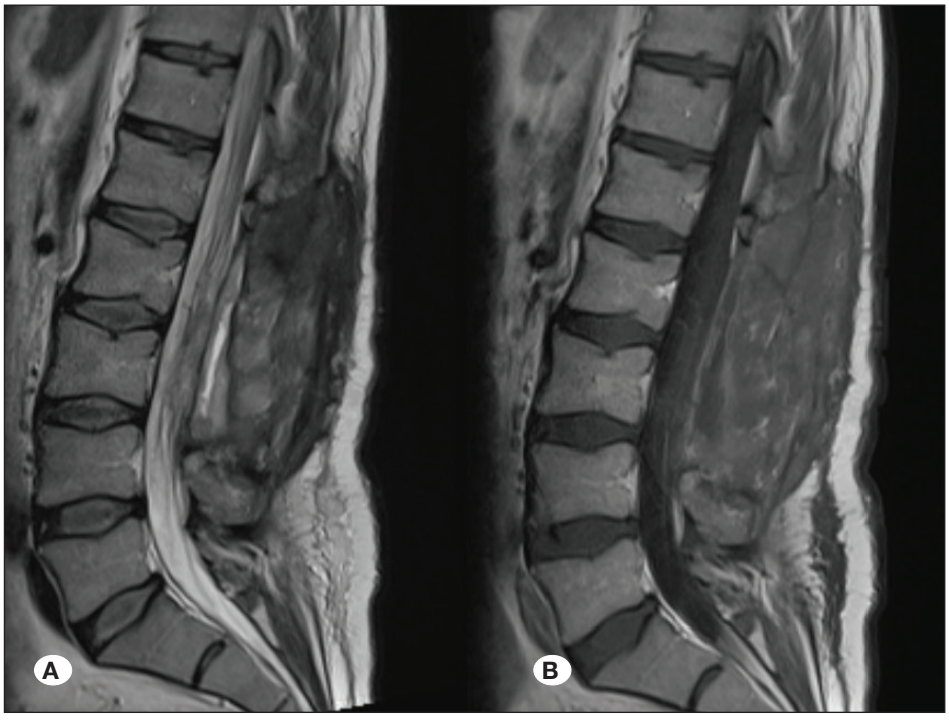

Figure 3: A) T2-weighted and **B)** T1-weighted imaging after surgery shows complete resection.

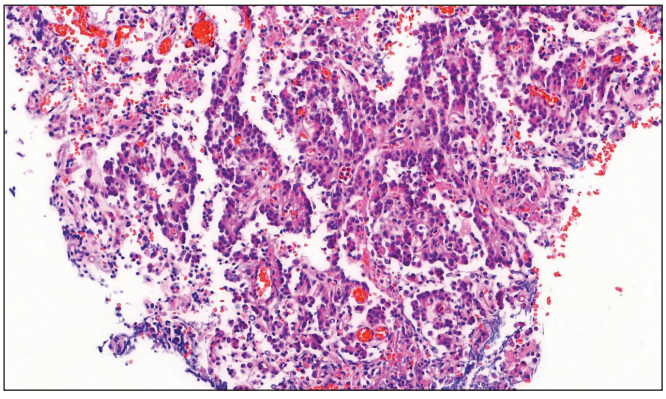

Figure 4: Papillary structures lined by low cuboidal cells (H&E, ×17.9).

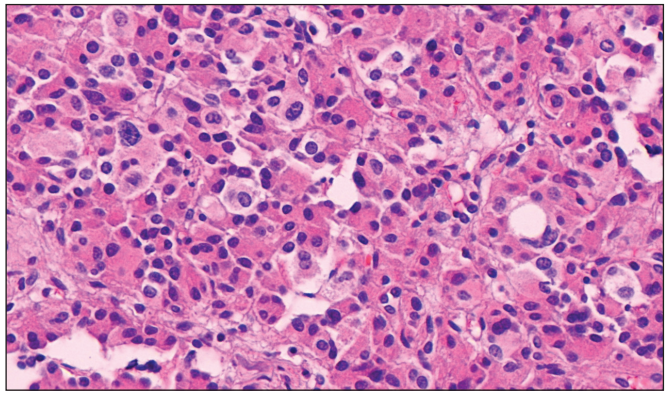

Figure 5: More solid area composed of mostly oncocytic tumor cells (H&E, ×46.7).

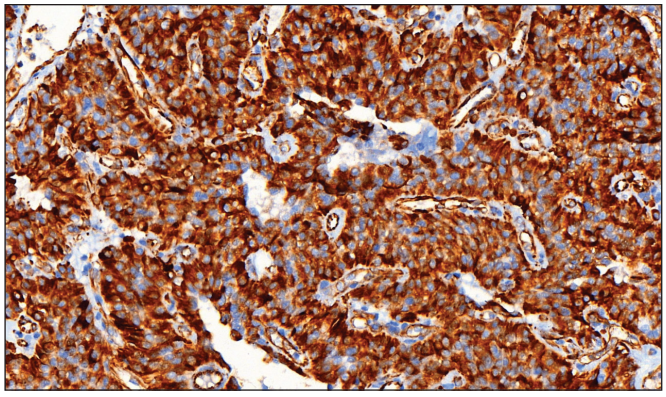

Figure 6: Diffusely strong vimentin expression in tumor cells (×28.2).

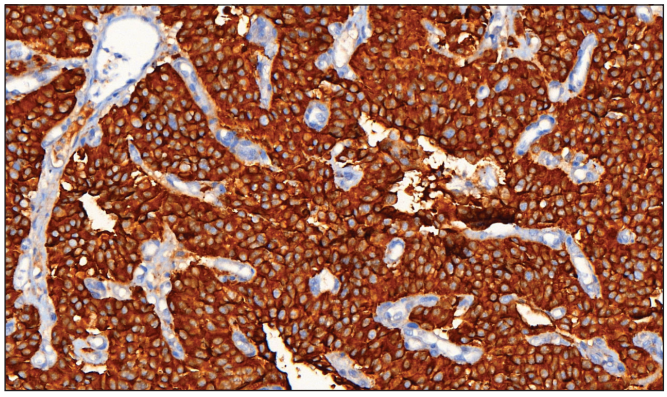

Figure 7: Diffusely strong synaptophysin expression in tumor cells (×27.4).

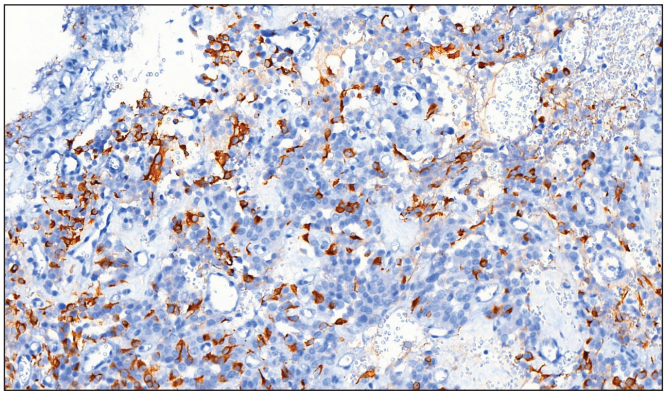

Figure 8: Cytokeratin expression in some tumor cells (×21.9).

The choroid plexus first appears in the fourth ventricle on the 41st day of embryologic development. Continuing with the ependyma, the choroid plexus epithelium develops from the neural tube. The leptomeningeal axis develops from the paraxial mesoderm. It is not clearly known when the choroid plexus begins to secrete cerebrospinal fluid. In normal embryological development, there is no choroid plexus in the spinal canal. Therefore, isolated choroid plexus neoplasia in the spinal region is not expected (9). Since the tumor in our case probably originated from choroid plexus cells that formed in the intradural spinal space during embryogenesis, it can be considered a choristoma. Development of a choroid plexus tumor in the spinal region can be attributed to defects in various stages of neuronal development, metaplasia of ependymal cells to choroid plexus cells, or ectopic neuro ectodermal residue. To prove these hypotheses, a better understanding of the embryogenesis stage and advanced immunohistochemical examination are required.

Katoh et al. reported an extraventricular atypical CPP and noted the importance of Kir7.1 immunohistochemical analysis in establishing the pathological diagnosis (6). Uff et al. reported a metastatic atypical CPP at the L5 level (12). There is a need for advanced immunohistochemical analysis in many extraventricular or spinal (synchronous or metastatic) tumors (2,3,8,10,11) . Such information may explain the pathogenesis of rare isolated spinal choroid plexus tumors.

CONCLUSION

To our knowledge, we have reported the first case of an isolated lumbar grade II atypical CPP. Unlike metastatic and synchronous tumors, the pathogenesis of isolated choroid plexus tumors within the spinal canal has not been explained. The importance of mitotic activity, immunohistochemical analysis, and tumor grade in isolated cases will be revealed in future studies.

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AUTHORSHIP CONTRIBUTION

Study conception and design: MCK

Data collection: MCK

Analysis and interpretation of results: MB

Draft manuscript preparation: MCK

Critical revision of the article: MB

Other (study supervision, fundings, materials, etc...): MCK

All authors (MCK, MB) reviewed the results and approved the final version of the manuscript.

REFERENCES

1. Boldorini R, Panzarasa G, Girardi P, Monga G: Primary choroid plexus papilloma of the sacral nerve roots. J Neurosurg Spine 10:51-53, 2009
2. Bollo RJ, Zagzag D: Samadaniu: Synchronous choroid plexus papilloma of the fourth ventricle and ependymoma of the filum terminal: Case report Neurosurg 67: E1454-E1459, 2010

3. Del Río-Pérez CM, Guillén-Quesada A, Alamar-Abril M, Candela-Canto S, García-Fructuoso G, Roussos I, Costa-Clara JM: Metastatic plexus papilloma in the pediatric age: A case report and review of the literature. Rev Neurol 54:673-676, 2012
4. Ikota H, Tanaka Y, Yokoo H, Nakazato Y: Clinicopathological and immunohistochemical study of 20 choroid plexus tumors: their histological diversity and the expression of markers useful for differentiation from metastatic cancer. Brain Tumor Pathol 28:215-221, 2011
5. Kaptanoglu E, Tun K, Celikmez RC, Ozen O, Taskin Y: Spinal drop metastasis of choroid plexus papilloma. J Clin Neurosci 14:381-383, 2007
6. Katoh M, Sawamura Y, Tanaka S, Fujimaki T, Hirose S, Aida T: Immunohistochemical analysis of cerebral intraparenchymal choroid plexus tumor: Case report. J Neurol Surg A Cent Eur Neurosurg 80(1):053-057, 2019
7. Kurtkaya-Yapicier O, Scheithauer BW, Van Peteghem KPV, Sawicki JE: Unusual case of extradural choroid plexus papilloma of the sacral canal. Case report. J Neurosurg 97 Supplement:102-105, 2002
8. Ochoa-Cacique D, Córdoba-Mosqueda M, Aguilar-Calderón JR, Sánchez-Silva MC, Vicuña-González RM, Ibarra-de la Torre A, Reyes-Rodríguez VA, de Jesús Lomelí-Ramírez J, Medina-Carrillo Ó, Sánchez-Calderón MD, Castañeda-Ramírez EA: Spinal drop of atypical choroid plexus papilloma in an adult patient: A case report and literature review. Surg Neurol Int 11:409, 2020
9. Sakka L, Coll G, Chazal J: Anatomy and physiology of cerebrospinal fluid. Eur Ann Otorhinolaryngol Head Neck Dis 128:309-316, 2011
10. Shi YZ, Chen MZ, Huang W, Guo LL, Chen X, Kong D, Zhuang YY, Xu YM, Zhang RR, Bo GJ, Wang ZQ: Atypical choroid plexus papilloma: Clinicopathological and neuroradiological features. Acta Radiol 58:983-990, 2017
11. Stuivenvolt M, Mandl E, Verheul J, Fleischeuer R, Tijssen CC: Atypical transformation in sacral drop metastasis from posterior fossa choroid plexus papilloma. BMJ Case Rep 2012, 2012
12. Uff CE, Galloway M, Bradford R: Metastatic atypical choroid plexus papilloma: A case report. J Neurooncol 82:69-74, 2007