Atypical choroid plexus papilloma: spontaneous resolution of diffuse leptomeningeal contrast enhancement after primary tumor removal in 2 pediatric cases

Marcello Scala, MD; Giovanni Morana, MD, PhD; Claudia Milanaccio, MD; Marco Pavanello, MD; Paolo Nozza, MD; and Maria Luisa Garrè, MD

Neuro-Oncology Unit, Neuroradiology Unit, and Departments of Neurosurgery and Pathology, Istituto Giannina Gaslini, Genova, Italy

Atypical choroid plexus papillomas can metastasize in the form of leptomeningeal seeding. Postoperative chemotherapy is the recommended first-line treatment when gross-total removal is not achieved or in cases of disseminated disease. Here the authors report on 2 children with atypical choroid plexus papillomas and MRI findings of diffuse leptomeningeal enhancement at diagnosis, later presenting with spontaneous resolution of the leptomeningeal involvement after removal of the primary lesions. Observations in this report expand our knowledge about the natural history and biological behavior of these tumors and highlight the role of close neuroimaging surveillance in the management of atypical choroid plexus papillomas in cases with MRI evidence of diffuse leptomeningeal enhancement at presentation.

https://thejns.org/doi/abs/10.3171/2017.2.PEDS16526

KEY WORDS: brain tumor; pediatric; atypical choroid plexus papilloma; leptomeningeal dissemination; oncology
Operation

The tumor was surgically treated via interhemispheric transcallosal approach. However, because of intraoperative bleeding, tumor removal was limited to a partial resection. Both lumbar and ventricular cerebrospinal fluid (CSF) collections were negative for neoplastic cells.

Pathological Findings

Neuropathological examination showed a papillary CPT with large areas of necrosis, without nuclear pleomorphism. Three mitoses per 10 randomly selected high-power fields were found. The Ki-67 proliferation index was 9%. Brain invasion was not found. The diagnosis was ACPP.

Second Operation and Postoperative Course

Two weeks after the initial resection, residual tumor was completely removed via a left frontal transcortical approach. One week later, postoperative MRI was performed as staging before the start of chemotherapy, which was considered given the presence of leptomeningeal enhancement on the preoperative scan. Magnetic resonance imaging confirmed gross-total removal and showed a reduction in leptomeningeal intracranial involvement (Fig. 1D and E). Strict MRI follow-up was then planned and no chemotherapy treatment was adopted. One month later, MRI demonstrated almost complete resolution of the intracranial and spinal leptomeningeal enhancement (Fig. 1F–H), which was no longer visible on a subsequent MRI performed 5 months later. The patient is still disease free 26 months after surgery (Fig. 1I and J).

Case 2

History and Examination

An 11-month-old boy was referred to our hospital because of macrocephaly. Neurological examination and funduscopy were normal. Brain MRI showed a large, left intraventricular, polylolobulated contrast-enhancing mass originating from the choroid glomus and extending into the ipsilateral temporal and occipital horns (Fig. 2A–C). No parenchymal infiltration was evident. Diffuse leptomeningeal enhancement was demonstrated into the basal cisterns and along the brainstem.

Operation

A left temporoparietal craniectomy was performed and gross-total tumor removal was achieved. Cerebrospinal fluid collections from lumbar puncture and ventricles were negative.

Pathological Findings

Neuropathological examination showed a papillary CPT with small areas of necrosis, without nuclear pleomorphism. Three mitoses per 10 randomly selected high-power fields were found. The Ki-67 proliferation index was 7%. Brain invasion was not found.
Postoperative Course

Magnetic resonance imaging performed 3 weeks after surgery (Fig. 2D–F) did not show residual disease and demonstrated also almost complete resolution of the leptomeningeal enhancement, which was no longer visible on subsequent MRI performed 1 month later. However, surveillance neuroimaging performed at 12 months revealed a small, growing, contrast-enhancing lesion located along the posterior margin of the surgical cavity (Fig. 2G). The nodule was surgically removed. No neoplastic cells were found in the ventricular CSF samples. The diagnosis of ACPP was confirmed. Two months later, MRI showed no residual disease and no signs of leptomeningeal involvement. Chemotherapy was never administered and the patient is still disease free 43 months after the first surgery (Fig. 2H and I).

Discussion

Choroid plexus papillomas metastasize as intraparenchymal or intraventricular nodules, especially subarachnoid nodules, or less frequently as diffuse leptomeningeal seeding. Metastases at diagnosis in cases of ACPP are not uncommon and have been reported in 17% of cases according to the Choroid Plexus Tumor–Society of Pediatric Oncology–2000 (CPT-SIOP-2000) study of CPTs. In cases of metastases, local recurrence, or incomplete resection, chemotherapy is recommended and the pa-
Vanishing arachnoidal enhancement in plexus papilloma

J Neurosurg Pediatr Volume 20 • September 2017

was demonstrated before any surgical procedure. Indeed, leptomeningeal enhancement occurred probably as a result of the growth of a macroscopic component along the resection margins, without any MRI sign of arachnoidal involvement. After surgical removal of the lesion, the same follow-up strategy was adopted and no adjuvant therapy was started. No signs of local recurrence or dissemination appeared in subsequent MRI.

Regarding the nature of the leptomeningeal enhancement, it has been supposed that it could be the result of abnormal arachnoidal vessel permeability triggered by hormonal factors (such as vascular endothelial growth factor) secreted by the tumor or of real arachnoidal infiltration by a lining of neoplastic cells, depending on a critical concentration of growth factors secreted by the tumor. The removal of primary tumor and thus the lack of a triggering factor may explain the resolution of leptomeningeal enhancement. At the same time, we recognize that leptomeningeal enhancement does not necessarily mean malignant seeding since it has been described in cases of obstructive hydrocephalus (venous stasis) or of leptomeningeal irritation from postoperative subarachnoid bleeding in children with brain tumors. The latter does not apply in our patients since leptomeningeal enhancement was demonstrated before any surgical procedure.

Regardless of the true cause, MRI evaluation is standard in the determination of secondary dissemination in brain malignancies, and the neuro-oncological strategy depends on MRI data even without histological confirmation. In our cases, given the location of the leptomeningeal involvement (basal cisterns and spine), an additional surgical procedure would have been necessary to evaluate the nature of the contrast enhancement, exposing the children to additional risks; therefore, close MRI follow-up was the preferred strategy. Since postoperative chemotherapy is the recommended first-line treatment for ACPP in cases of disseminated disease, MRI evidence of diffuse leptomeningeal contrast enhancement at presentation in ACPP cases should be interpreted with caution, and restaging should be performed after gross-total removal or subtotal removal of the tumor. Indeed, leptomeningeal enhancement in the setting of ACPP may represent a tumor-related effect rather than true tumor dissemination. In conclusion, our observations expand our knowledge about the natural history and biological behavior of ACPP and highlight the role of close clinical and neuroimaging monitoring in the management of ACPP with MRI evidence of leptomeningeal involvement at presentation.

Acknowledgments

We acknowledge support from the Association for Pediatric Brain Tumors (Associazione per la ricerca sui tumori cerebrali del bambino) and the Berlucchi Foundation.

References


Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Scala, Morana, Garrè. Acquisition of data: Scala, Morana. Analysis and interpretation of data: Scala, Morana, Garrè. Drafting the article: Scala, Morana. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Scala. Study supervision: Morana, Garrè.

Correspondence
Marcello Scala, Neuro-Oncology Unit, Istituto Giannina Gaslini, Via Gerolamo Gaslini, 5, Genova 16148, Italy. email: marcelloscala87@gmail.com.