Case Reports

Case 1

History and Examination

An 8-month-old boy was admitted for the gradual onset of weight loss, poor feeding, downward gaze, and vomiting. At presentation, he was macrocephalic with bilateral papilledema. Brain MRI showed a huge cauliflower-like contrast-enhancing mass filling and expanding the third ventricle (Fig. 1A), without evidence of parenchymal invasion. There was concomitant hydrocephalus. Diffuse pathological contrast enhancement was present in the basal region.
cisterns, on the surface of both cerebral hemispheres, and along the spinal cord (Fig. 1A–C).

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 fundoscopy were normal. Brain MRI showed a large, left intraventricular, polylobulated 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-
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apply in our patients since leptomeningeal enhancement should be performed after gross-total removal or subtotal cases should be interpreted with caution, and restaging of disseminated disease, MRI evidence of diffuse leptomeningeal enhancement, a well-differentiated choroid plexus papilloma. Acta Neurochir (Wien) 144:723–728, 2002

References


Acknowledgments

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

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

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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.

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