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Pilocytic Astrocytoma of the Cerebellopontine Angle with cerebrospinal fluid Spread in an Adult: A Case Report

Astrocitoma pilocítico no ângulo pontocerebelar com disseminação liquórica em paciente adulto: Relato de caso

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Abstract

Introduction Pilocytic astrocytoma of the cerebellopontine angle (CPA) is uncommon, and its spread to the cerebrospinal fluid (CSF) at the time of diagnosis has not been reported in the literature.

Case Presentation We report the case of a 33-year-old man with multifocal pilocytic astrocytoma diagnosed by magnetic resonance imaging (MRI) and confirmed by histopathological examination, and present the radiological and histopathological findings.

Conclusion In the case herein reported, we observed spread of the pilocytic astrocytoma of the CPA to the CSF at the initial diagnosis, and early detection by MRI is very important regarding the treatment modality and prognosis.

Keywords
- cerebellopontine angle
- pilocytic astrocytoma
- vestibular schwannoma

Resumo

Introdução O astrocitoma pilocítico no ângulo pontocerebelar (APC) é incomum, e sua disseminação liquórica no momento do diagnóstico não foi relatada na literatura.

Apresentação do Caso Relatamos o caso de um homem de 33 anos com astrocitoma pilocítico multifocal diagnosticado por ressonância magnética (RM) e confirmado por exame histopatológico, e apresentamos os achados radiológicos e histopatológicos.

Conclusão No caso relatado, observou-se disseminação liquórica de astrocitoma pilocítico no APC no diagnóstico inicial, e a detecção precoce por RM é muito importante para a modalidade de tratamento e o prognóstico.

Palavras-chave
- ângulo pontocerebelar
- astrocitoma pilocítico
- schwannoma vestibular

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Introduction

Tumors of the cerebellopontine angle (CPA) comprise 6% to 10% of intracranial neoplasms. The most common CPA neoplasms are those of the vestibular schwannoma. Pilocytic astrocytomas (PAs) are low-grade astrocytomas that occur more often in children and young adults, mostly near the midline, usually arising from the cerebellum, the optic chiasm, and the hypothalamic region. They are seldom seen within the CPA.[1] In the literature, they are often the exophytic extension of the primary brainstem or cerebellar PAs at this location. Reports of primary extra-axial PA are extremely rare. Primary spread to the cerebrospinal fluid (CSF) is seen mostly observed in cases of ependymoma, germ-cell tumors, and high-grade glial tumors**.** According to the classification pf the World Health Organization (WHO), PAs are classified as grade-1 gliomas. However, though uncommon, PAs may spread to the CSF.[2]

We present a case of CSF spread of a PA of the CPA in an adult.

Case Presentation

A 33**-**year-old male patient was admitted to our neurosurgery department with complaints of dizziness, tinnitus, and mild headache that had started in the previous year. His medical and family histories were non-contributory. Upon physical examination, he was normotensive and his distal upper extremity pulse and heart rate were normal. The presence of neurofibromatosis stigma was not observed. No abnormal finding was revealed in the neurological examination. The routine laboratory findings were normal. The hematologic tests were negative for all rheumatologic and autoimmune disorders.

A brain magnetic resonance imaging (MRI) scan revealed a lesion measuring $5 \times 3 \times 2.5$ cm in the left CPA, which was predominantly hypointense on T1 and hyperintense on T2-fluid-attenuated inversion recovery (FLAIR) weighted images. A CSF cleft between the mass and the brainstem suggested an extra-axial origin of the tumor. The mass was characterized by heterogeneous patchy enhancement and composed of both solid and cystic components (►**Fig. 1**). There was mild mass effect on the left side of the brainstem structure and on the fourth ventricle, without apparent signal abnormality. No diffusion restriction and peritumoral edema were detected. An axial three-dimensional (3D) constructive interference in steady state (CISS) sequence revealed a large hypointense filling defect within the left CPA with extension into the widened left internal auditory canal (IAC). Upon the initial examination, there were additional tumors located in the right Meckel cave and prepontine cistern, but no enhancement was observed in these lesions (►**Fig. 1**).

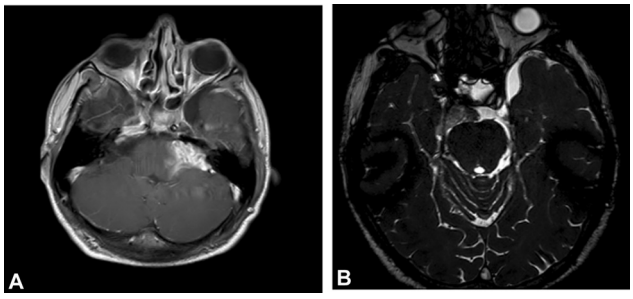

Fig. 1 Axial gadolinium-enhanced T1 weighted image (**A**) showing a patchy contrast-enhanced lesion on the left CPA with mild mass effect of the adjacent brainstem structure. Axial 3D CISS sequence (**B**) showing a large hypointense mass in the prepontine cistern.

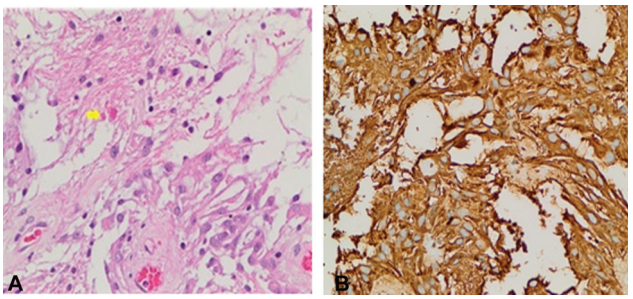

Fig. 2 Perivascular arrangement of elongated bipolar glial cells and microcyst and eosinophilic granular bodies (yellow arrow) in fibrillary background (**A**) (hematoxylin and eosin staining, 20× original magnification). Strong cytoplasmic GFAP immunoreactivity in neoplastic glial cells (**B**) (GFAP immunostaining, 40× original magnification).

The patient subsequently underwent surgical resection via the left suboccipital retrosigmoid approach. A gelatinous, yellowish-white, encapsulated, and normovascular tumor was identified in the CPA. Subtotal resection was performed because the tumor was attached to the left facial and vestibulocochlear nerves.

The histopathological evaluation showed a glial tumor composed of bipolar elongated neoplastic glial cells arranged around perivascular areas, resembling pseudorosettes. There were mulberry-shaped eosinophilic granular bodies and loose myxoid microcystic areas in the fibrillary background. Microvascular proliferation, mitosis, necrosis and microcalcification were absent. Immunohistochemical staining revealed immunopositivity for glial fibrillary acidic protein (GFAP), Olig2 and S100, and immunonegativity for epithelial membrane antigen (EMA) (►**Fig. 2**). The Ki 67 proliferative index was lower than 2%. A final diagnosis of PA (WHO grade I) was made. But the remote lesions were not histologically confirmed as PAs.

The patient was discharged after the surgery and recovered without any significant complications. Adjuvant irradiation treatment and regular follow-up with brain MRI scans were recommended because of the residual tumor mass in the CPA.

Discussion

Gliomas of the CPA are rare entities in adults, and are often the exophytic extension of the primary brainstem or of cerebellar gliomas.[3] In the case herein reported, the primary lesion was located at the CPA and did not have any intra-axial component on preoperative imaging. Intraoperatively, these findings were confirmed by the absence of adherence of the tumor capsule to the brainstem or cerebellum and by the presence of a clear boundary between them.

To date, less than 50 cases of primary extra-axial CPA gliomas have been reported in the literature. Although PAs are low-grade tumors, they have been documented to spread to other parts of central nervous system. Spread to the CSF of a PA of the CPA at the time of diagnosis (the case herein reported) is the first such case reported in the literature.

Preoperative neuroimaging studies suggested the diagnosis of cystic CPA schwannoma with the involvement of the IAC.

In the literature, spread has been defined as a leptomeningeal spread or a tumor found at sites other than that of the primary disease location on MRI. Primary CSF dissemination is mostly observed in cases of ependymoma, germ-cell tumors, and high-grade glial tumors. The spread of Pas to the CSF is uncommon and usually occurs after interventional procedures. The detection of spread during the primary diagnosis, as in the case herein reported, is extremely rare. The mechanism of the CSF spread of the PA is not clear. According to the literature, PA spread to the CSF is facilitated by its proximity to locations such as the ventricular system and basal cisterns. Although this type of spread usually follows the normal CSF flow, in the present case it was in the opposite direction. Mamelak et al.[4] reported that hypothalamic tumors are more prone to spread. But in the present case, the primary tumor was located in the CPA. The case reported by Dutta et al.[1] is similar to the one herein reported because of the size and imaging findings of the primary tumor, but it differs from our case due to the absence of evidence of CSF spread.

The origin of extra-axial gliomas of the CPA is still uncertain. Several theories have been suggested. One theory is that gliomas in this location arise from adjacent anatomical structures, such as the medial velum of the lateral recess of fourth ventricle. Another theory is that the tumor originates primarily in the transformation of heterotopic neuroglial cell nests in the leptomeninges covering the proximal cranial nerves.[1,3,5] In the current case, there was no history of intraparenchymal glioma and no intraoperative evidence of an anatomic association to the lateral recess of the fourth ventricle. We believe that the tumor most probably originates from neuroglial cells.

Conclusion

Pilocytic astrocytoma with CSF spread is a rare entity; therefore, the appropriate therapy for these patients is poorly defined. Mamelak et al. showed that surgery of the primary tumor is the preferred treatment modality for PAs that present spread. It has been documented that total surgical resection results in satisfactory tumor treatment with excellent outcomes and minor morbidity.[4] Pilocytic astrocytoma should be kept in mind in the differential diagnosis of unusual CPA tumors with atypical imaging findings. The possibility of spread of these tumors to another location should always be considered, and, after the primary treatment, radiological follow-up should be performed for the early diagnosis of the spread.

Consent to Participate

The patient herein described has given written consent regarding the inclusion of material pertaining to his case. The patient acknowledges that he cannot be identified through the paper. The authors have fully anonymized the patient.

Consent for Publication

The patient signed an informed consent form regarding the publication of his data.

Availability of Data and Material

The datasets analyzed in the current study are available from the corresponding author upon reasonable request.

Author's Contribution

Dr. Biyikli, Dr. Kursun, and Dr. Bozkurt conceptualized and designed the study, drafted the initial manuscript, and reviewed and revised the final manuscript. Dr. Biyikli, Dr. Kursun, Dr. Oguzsoy, Dr. Bozkurt, and Dr. Bayrakli designed the data collection instruments, collected data, carried out the initial analyses, and revised and critically reviewed the manuscript for important intellectual content. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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Conflict of Interests

The authors have no conflict of interests to declare.

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