CASE REPORT

Primary diffuse choroid plexus T-cell lymphoma: Case report

Lymphome diffus primitif à cellules T des plexus choroïdes

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KEYWORDS
Cerebral ventricles; Choroid plexus; Lymphoma; T-cell

Summary We present here a case of primary choroid plexus T-cell lymphoma with no evidence of immunodeficiency or immunological disease. As ventricular T-cell lymphoma is extremely rare, there is only limited information on the radiological findings of ventricular T-cell lymphoma. In this report, we also include some unusual MRI findings in this case that have never been described before.

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Introduction

Intraventricular neoplasms are uncommon, representing only 1–10% of all central nervous system (CNS) tumors [1]. Approximately 2.3% of all intracranial tumors are primary CNS lymphomas (PCNSL), although the incidence is higher in immunosuppressed patients [2,3]. However, only 1–3.6% of all lymphomas are T-cell CNS lymphomas (TCNSL). Involvement of the choroid plexus by malignant lymphoma is rare. Furthermore, to the best of our knowledge, only a few cases of primary diffuse choroid plexus TCNSL have been reported in the literature [2].

Case report

A 63-year-old woman presented with postictal unconsciousness. It was the first seizure in her entire life, and she had been complaining of progressively worsening headaches and dizziness for one week. Her medical history contained no evidence of immunodeficiency, nor were there any clinical or laboratory signs of immunological involvement. As computed tomography (CT) had revealed enhanced masses within all ventricles along with tetraventricular hydrocephaly, magnetic resonance imaging (MRI) was also
performed. Fluid attenuated inversion recovery (FLAIR) images revealed high signal intensity around the ventricles suggestive of periventricular edema. T2-weighted MR images showed marked enlargement of reduced intensity of the choroid plexus, while post-contrast T1-weighted MR images showed an intraventricular mass located within the choroid plexus. The lesion extended into the fourth ventricle via the third. Also, the lateral ventricles showed diffuse ependymal enhancement (Fig. 1).

The patient underwent an urgent operation involving suboccipital craniotomy with a navigation-assisted transvermian approach. Cerebrospinal fluid (CSF) drainage was performed, and a biopsy to determine the pathology was taken. The lesion was soft in nature and yellowish in color, arising from the choroid plexus and floating inside the ventricle. The histological diagnosis was high-grade diffuse large T-cell lymphoma (Fig. 2).

Whole-body CT scan showed no systemic involvement, and bone-marrow biopsy was normal. Cervical, thoracic and lumbar spinal MRI was also normal. A diagnosis of primary TCNSL was formulated.

Discussion

More than 90–95% of cases of PCNSL are histologically classified as high-grade diffuse large B-cell lymphoma; the remaining 5–10% includes T-cell lymphoma, Burkitt’s lymphoma, low-grade PCNSL, lymphomatosis cerebri and marginal zone B-cell lymphoma of the dura. PCNSL lesions may be classified as parenchymal, subependymal and leptomeningeal, with a possibility of ventricular involvement (2–8.6% of cases) that is nearly always secondary to extension from a subependymal location in a large number of cases. Primary ventricular lymphomas are extremely rare [1,4,5].

The choroid plexus is a tulle-like pinkish structure that floats in the CSF of the ventricles and attaches to the ependyma by a thin stalk. Tumors arising from it are extremely rare, and include choroid plexus papilloma, choroid plexus carcinoma and non-neoplastic lesions. Of these, lymphoma is an extremely rare form of choroid plexus lesion [1,2,6].

Because of its rarity, only a few series include TCNSL in the literature. Shenkier et al. [7] reported on a retrospective combined series of patients with TCNSL from 12 cancer centers in seven countries. The cerebral hemisphere was the most common primary site in all reported series. Other primary sites include the cerebellum, basal ganglia, corpus callosum and brain stem, accounting for around 10% of all cases. The meninges and spinal cord are unusual sites, accounting for 2–4% of cases. T-cell lymphoma of the choroid plexus is extremely rare, and usually arises from a systemic disease at a rate of 0.9% of cases [8,9].

Figure 1  A: high-intensity periventricular white-matter changes suggest edema on an axial FLAIR image; B: an axial T2-weighted image reveals enlargement of the choroid plexus and a low signal intensity lesion in the choroid plexus. Post-contrast axial (C), sagittal (D) and coronal (E) T1-weighted images show smooth linear enhancement of the ventricular walls. Infiltration of the choroid plexus is evident, as is extension of the lesion. The lesion emanating from the choroid plexus has grown into the fourth ventricle via the third ventricle. All of the ventricles are filled with the mass.
Involvement of the choroid plexus by lymphoma remains a diagnostic challenge to distinguish these lesions from non-neoplastic and other benign tumors. Ventricular lymphoma commonly arises in adults and does not originate from the ventricular atrium. Iso- to low T1-weighted and iso- to slightly high T2-weighted signal intensities and rim enhancement on MRI have been described [9]. Choroid plexus papilloma is a predominantly solid tumor with a homogeneous intensity that is well defined. It may also present as cysts with a mural nodule, and should be considered in the differential diagnosis along with hemangioblastoma and glioma. In addition, the differential diagnosis should include ependymoma, which presents with a ring-like CSF signal around the tumor, while the mass has an obvious border [10].

Park et al. [4] reported on a ventriculitis-type cyst-like intraventricular lymphoma, with a wall that was seen as high intensity in T1-weighted images with gadolinium enhancement. Our present case also showed enhancement of the ependymal surface mimicking ventriculitis.

Terasaki et al. [2] described a case of primary choroid plexus T-cell lymphoma, where the lesion showed rim enhancement in the choroid plexus. Our present case showed diffuse enhancement of the whole of the choroidal structures within all of the ventricles.

In conclusion, T-cell lymphoma of the ventricle is extremely rare and may present with various radiological findings. In patients with an intraventricular mass and diffuse ependymal enhancement, a differential diagnosis of a possible intraventricular lymphoma should be considered.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.