Somalia Türkiye Medical Journal (STMEDJ)
V. 02 No. 01, 2023
doi:10.58322/stmj.v2i1.16
http://www.stmedj.com
eISSN: 2958-2849

Case Report

Central Neurocytoma Presenting with Progressive Headache: A Case Report and Literature Review

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Received November 20th, 2022
Accepted December 22nd, 2022

Abstract Central neurocytoma (CN) is a rare benign brain tumor primarily diagnosed in young patients. It mainly occupies the lateral ventricle and has relatively characteristic imaging features that aid in its diagnosis. This tumor causes obstructive hydrocephalus and manifests with signs of increased intracranial pressure. Herein, we present a case of a 53-year-old female diabetic patient who presented to our neurology clinic with an eight-month history of diffuse progressive headaches. Her headache was associated with nausea but not vomiting. There was no motor or sensory deficits or paresthesia. Likewise, the examination revealed no diplopia, ophthalmoparesis, cranial nerve impairments, or papilledema. Magnetic resonance imaging (MRI) of the brain revealed a heterogeneous intraventricular mass measuring 45x41 mm in size with surrounding mild tissue edema, typical for a central neurocytoma. The patient was referred to the neurosurgery department for surgical removal of the tumor; however, the patient did not consent to surgery. Brain MRI should be done in young patients with a headache that does not have the characteristics of primary headaches.

Keywords Central neurocytoma, Brain tumor, Progressive headache.

Introduction

Central neurocytoma is an uncommon intraventricular benign central nervous system neoplasm that accounts for 0.25–0.5% of primary brain tumors. It is relatively more common in younger patients; 70% of the cases are diagnosed between the ages of 20 and 40 (**1**). Although some of these tumors have been

reported to be located in the third and fourth ventricles, most are located in the anterior portion of the lateral ventricle. In addition, the tumor is often connected to the septum pellucidum at the foramen of Monroe (**2**). Central neurocytoma can elevate intracranial pressure and thus induce hydrocephalus. Nausea, vomiting, headaches, seizures, loss of consciousness, weakness, and memory or vision impairment are all possible symptoms or signs. However, the duration of symptoms appears to be primarily related to the tumor location rather than the tumor's aggressiveness (**3**). The diagnosis is made through brain MRI, but the definitive diagnosis is made by histopathology (**4**).

Here, we report a patient with lateral ventricle neurocytoma (a rare brain tumor) presenting with a chronic progressive, moderately severe headache with no other associated neurologic deficit.

Case Presentation

A 53-year-old female diabetic patient came to the neurology clinic with an eight-month history of diffuse and progressive headaches. The headache was more on the vertex and was of moderate intensity. It was associated with nausea but not vomiting. There were no associated photophobia, phonophobia, or change in the headache in different head positions.

The patient did not have a family history of brain tumors. The patient had no paresthesia or motor or sensory impairments upon neurologic assessment. The examination revealed no signs of diplopia, ophthalmoparesis, or cranial nerve impairments. The fundus examination revealed no associated papilledema, and the rest of the neurologic examinations were unremarkable.

Brain MRI revealed a heterogeneous intraventricular mass lesion 45x41 mm in size and surrounding mild tissue edema, making a compression effect on the right lateral ventricle (**Figures 1-4**). The location and signal characteristics of the lesion were consistent with central neurocytoma. Unfortunately, histopathology and MRI spectroscopy could not be performed due to a lack of availability. The patient was referred to the neurosurgery department for surgical treatment. Surgical removal was recommended; however, the patient did not consent to surgery.

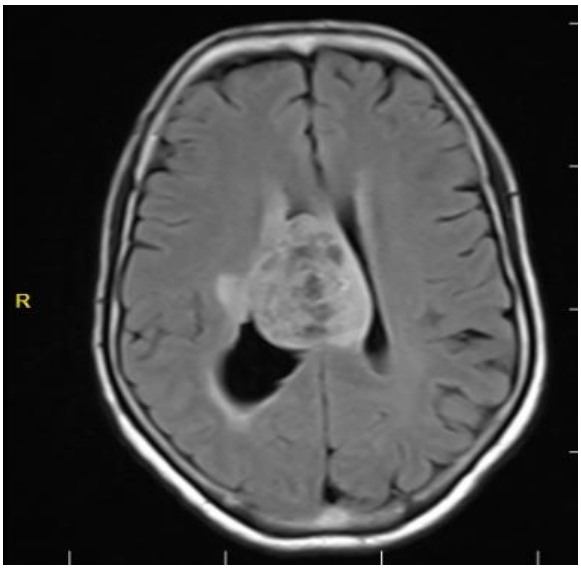

Fig.1. Brain MRI Axial Flair view, showing hyperintense heterogeneous intraventricular lesion with mild edema

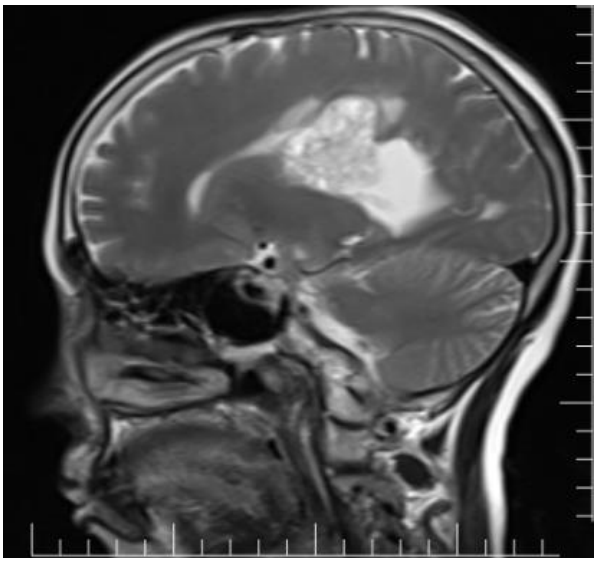

Fig.2. Brain MRI T2 Sagittal view showing hyperintense intraventricular mass lesion with mild

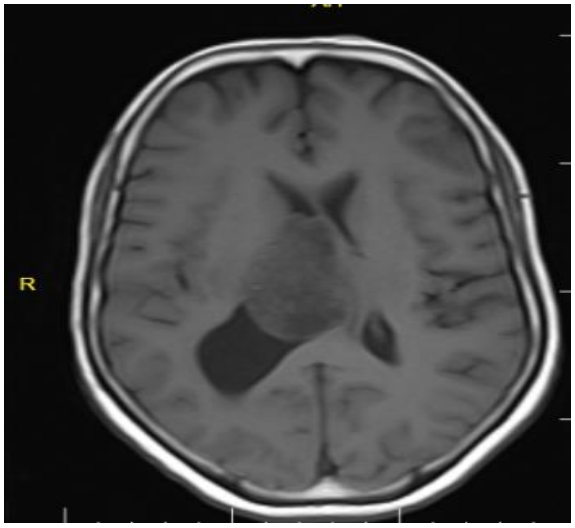

Fig.3. Brain MRI T1 Axial view showing isointense intraventricular mass lesion with mild lateral ventricular compression.

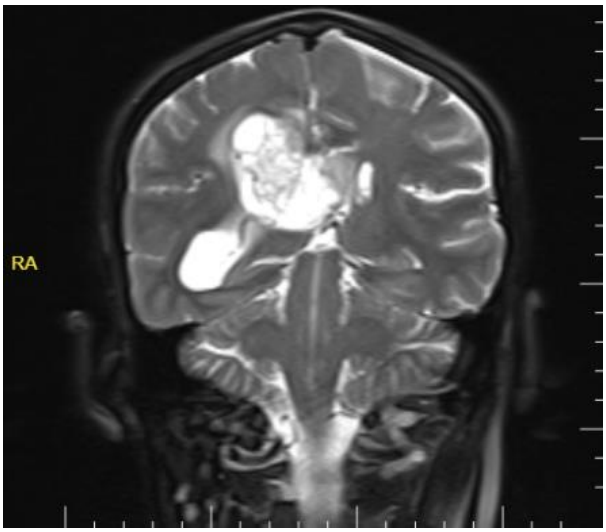

Fig.4. Brain MRI T2 Coronal View showing hyperintense intraventricular mass lesion with mild edema around it and lateral ventricular compression.

Discussion

Central neurocytoma is a rare benign central nervous system tumor that usually develops in the lateral ventricles. Many advances have been made in diagnosis and therapy since it was first reported in the early 1980s(**4**). Despite the advances, identifying the causes of these rare tumors and developing effective new treatment techniques remains challenging. Central neurocytoma accounts for 0.1–0.5% of all primary brain tumors. Because CN is usually intraventricular, it manifests clinically as hydrocephalus (**5**). Although CN is more common in teenagers and young adults, it has also been documented in patients who are elderly or very young (**2**). Hydrocephalus is the common symptom of most neurocytomas, which is caused by the obstruction of the foramen of Monroe by the tumor. Increased intracranial pressure causes symptoms such as headaches, visual impairment, nausea, and vomiting. Other signs and symptoms include weakness, dizziness, paresthesia, tinnitus, seizures, memory loss, and loss of consciousness. Papilledema and ataxia are two common symptoms (**5**, **6**). Our patient had a moderately severe headache that had worsened over the last eight months. The headache was not associated with nausea, vomiting, motor/sensory deficits, or paresthesia during the test. The examination revealed no diplopia, ophthalmoparesis, or cranial nerve impairments, as well as no concomitant papilledema on the fundus examination.

Diagnosis is generally put by the combination of MRI with immunohistochemical confirmation of specific molecular markers. Central neurocytomas comprise lobulated masses with well-defined borders and cyst-like areas (**7**). In magnetic resonance imaging (MRI), central neurocytomas are isointense to gray matter on T1-weighted sequences and hyperintense on T2-weighted images. These lesions may appear "bubbly" due to numerous cysts (**8**, **9**). In our case, a brain MRI revealed a 45x41 mm heterogeneous intraventricular mass lesion with slight edema around it, which is typical of CN. Tumors in the lateral ventricle in young adults include subependymal hamartomas, which may look similar. They are typically found in the fourth ventricle and are usually more common in older individuals. They present with obstructive hydrocephalus, which is lacking in this case. Central neurocytoma in the lateral ventricle is usually adjacent to the septum pellucidum as one of its distinctive features.

Central neurocytoma shares many of the same histopathologic features as oligodendrogliomas and ependymomas. As a result, they have been misdiagnosed in the past. Immunohistochemistry and electron microscopy are used to validate the neuropathic diagnosis of CN (**7**). Unfortunately, due to a lack of resources, histopathology and immunohistochemistry could not be performed. Therefore, the diagnosis was based on the clinical picture and radiological data.

The goal of treatment is to achieve a gross total resection (GTR), which has an excellent prognosis and effective tumor control. The primary goal of the surgery is to achieve maximum resection with minimal neurologic compromise, restore the CSF pathways, and provide tissue for definitive histopathological diagnosis. Because most CNs are intraventricular and do not infiltrate the surrounding parenchyma, microsurgical techniques can be performed during gross total resection (**1**, **9**). The GTR can be successfully performed in about half of the patients. Tumor size, location, lesion extent, attachment to surrounding structures, vascularity, and surgeon experience play essential roles in resectability. When GTR is impossible, adjuvant radiosurgery and radiotherapy may be considered to improve tumor control (**10**, **11**). We referred our patient to the neurosurgery department for surgery. Surgical removal was recommended, but unfortunately, the patient and her family did not accept this treatment.

Conclusion

Central neurocytoma is a rare benign brain tumor presenting with signs of increased intracranial pressure. Brain MRI should be done in young patients with a headache that does not have the characteristics of primary headaches.

Consent for publication: Written informed consent was obtained from the patient to publish this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Availability of data and materials: The datasets used and analyzed during the current study are available from the corresponding author upon reasonable request.

Funding: This study was unfunded.

Conflict of interest: The authors declare no conflicts of interest.

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