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Central Neurocytoma: Case Report and Review of Literature

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Abstract

Central neurocytoma is a rare intra-ventricular brain tumor that affects young adults and presents with increased intracranial pressure secondary to obstructive hydrocephalus. Usually, it has a good prognosis after sufficient surgical intervention, but in some patients the clinical course is more invasive. In this report, we report a case of childhood central neurocytoma with focusing on incidence and chemotherapy treatment at our oncology center.

Keywords: Central neurocytoma, Pediatric, Brain tumor.

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Introduction

Central neurocytoma is an extremely rare benign tumor that arise most of the times in the lateral ventricles near the Monro foramina (0.1 - 0.5% of all primary brain tumors). Also this entity is rarer in pediatrics compared with adult. It was first explained in 1982 by Hassoun and ws arranged as WHO grade II tumors [1,2,3].

Accurate Incidence for central neurocytomas are not clear. Male to female ratio of 1:4, but it is now believed that both sexes are equally affected. No specific geographic distribution has been reported. Risk factors have not been identified [4,5,6]. We didn't find incidence rate in children purely Central neurocytoma till 2016 has 500 known cases [7].

Origination of tumor from lateral ventricles leads to blurred vision and increased intracranial pressure [8].

The standard treatment for a central neurocytoma is surgical resection in pediatrics and adult. Typically there are no mitoses, nucleoli, or infiltrating margins. Immunohistochemically (IHC) these tumors are highly positive for neuronal differentiation (synaptophysin, neuron specific enolase), focal glial fibrillary acidic protein (GFAP) and negative for oligodendrocyte transcription factor [9,10].

The differential diagnosis are ependymoma, astrocytoma, intra-ventricular oligodendrogliomas or primary cerebral neuroblastoma but definite diagnosis is made by immunohistochemical methods [11].

Case Presentation

The patient was a 3.5-year-old Afganian boy resident in Fars province in Iran was admitted to Namazi hospital in Shiraz with headache, nausea and vomiting that had lasted for 21 days before admission (Figure 1).

In physical examination, bilateral papilledema was noted, without any neurological deficits. Brain computed tomography (CT) scan showed a large, heterogeneous mass in third and left lateral ventricles with hydrocephalus and small vasogenic edema surrounding the lesion. Treatment

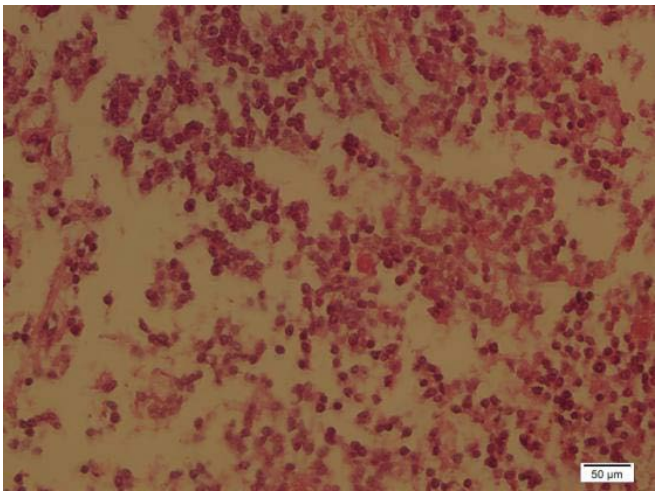

Figure 1. *Histopathology of the lesion resected during the craniotomy of a 3.5-year-old male. Hematoxylin and eosin staining indicates a tumor of moderate cellularity with vascular proliferation, as indicated by the arrow (magnification, ×400).*

with manitol 20% (1 gr/kg) and Dexamethasone (0.15 mg/kg q 6 h) was prescribed for decreasing intracranial pressure and cerebral edema (Figure 2).

Magnetic resonance imaging (MRI) showed a heterogeneous mass 35 × 41× 48 mm in size in mentioned places with solid component. There was a mass effect on the lateral ventricle, with midline shift of about 6mm from right frontal cortex. Complete spinal imaging was performed using MRI, with no evidence of drop metastasis and subarachnoid dissemination of the tumor being identified. The patient underwent right craniotomy for tumor resection. His tumor was done biopsy with neurosurgeon (Figure 3).

Histologically, the tumor exhibited uniform small round cells with regular nuclear morphology. In Immunohistochemical study (IHC), the tumor cells showed perinuclear strong positivity for synaptophysin and focal positivity for oligodendrocyte transcription factor 2.

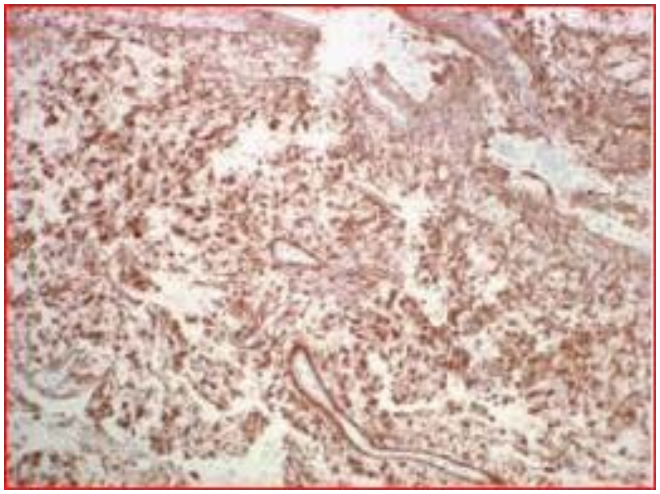

Figure 2: *Synaptophysin immunostain showing strong positive staining, indicating the neuronal origin of the tumor*

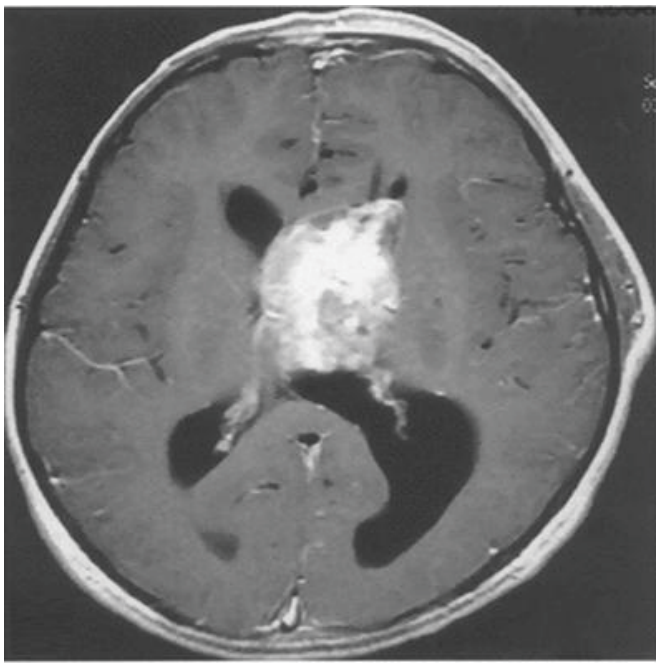

Figure 3: *Preoperative MRI (Gd-enhanced T1). A heterogeneous mass 35X41X48 mm in size was observed in third and left lateral ventricle with hydrocephalus.*

In addition, strong immunopositivity for vimentin and CD99 was noted. The MIB-1 (Ki-67) was 30%. The tumor wasn't resected completely. Radiotherapy was refused by the parents due to the patient's young age, but they accepted chemotherapy. After surgery, the patient doesn't recover full consciousness.

He received first course of chemotherapy including Etoposide, 40 mg/m^2/day and Cisplatin 25 mg/m^2/day for 4 days and Cyclophosphamide 1,000 mg/m^2 on Day 4. After receiving this regimen his parents released and discharged him from hospital and discontinued treatment.

Discussion

To this date, limited information about central neurocytoma has been reported in the pediatric population.

This tumor usually affects young adults and is located in the lateral and third ventricles, causing obstruction to the liquor drainage – headaches and other symptoms of raised intracranial pressure [12].

Our patient similar other typical neurocytoma cases presented with intracranial mass symptoms during a short time. This tumor can present with other clinical features such as obstructive hydrocephalus due to intraventricular hemorrhage within the tumor, chronic progressive blurring of vision, gigantism, Behavioral disorders, also visual and auditory hallucinations were reported [13-17].

The treatment for neurocytoma in pediatric and adult populations is surgical resection, with post-operative radiation therapy for incomplete resection or recurrence. In addition, drop metastasis must be intently eluded during resection surgery [18]. Our patient hadn't any evidence of spinal involvement.

Despite of benign pathology in majority of patients diffused infiltration of the brainstem, cerebellum and spinal cord during primary setting or relapse was reported [19,20].

Also, peritoneal dissemination from central neurocytoma especially in patients with ventriculoperitoneal shunt was seen therefore evaluation of proliferative index may be a guideline parameter for planning adjuvant therapies after surgical treatment in selected cases [21].

Radiotherapy is another option used as adjuvant treatment when resection is incomplete or when central neurocytoma is a hyper vascular tumor [22]. Sometimes Gamma Knife stereotactic radiosurgery for residual or recurrent tumors especially at early detection before tumor progression is counseled [23].

Behavioral disorders were seen as an unusual presentation of pediatric extraventricular neurocytoma also visual and auditory hallucinations were reported [24,25].

Although chemotherapy role is not well proved for patients with central neurocytoma, it has been used as an adjuvant treatment when the tumor resection is incomplete. In our patient, after surgery and incomplete resection chemotherapy it was prescribed.

In the literature administration of vincristine and carboplatin for 6 of 8 total cycles after 18 months follow-up and serial imaging, shows stable disease within the craniospinal axis in pediatric patients but for our patient craniospinal axis imaging wasn't done [26].

The use of chemotherapy is controversy in kids. Intensive chemotherapy followed by autologous stem cell rescue for atypical neurocytoma may be considered as an adjunct to surgical therapy in young patients with atypical neurocytoma not amenable to radiation therapy [27]. Six cycles of chemotherapy including Procarbazine, CCNU, and Vincristine previously described for the treatment of a central neurocytoma and presents an additional treatment option [28] comprised of Etoposide, 40 mg/m^2/day, for 4 days; Cisplatin, 25 mg/m^2/day, for 4 days; and cyclophosphamide, 1000 mg/m^2, on Day 4; this cycle was repeated every 4 weeks led to long term disease reduction [29]. In our patient because of refusing treatment continuation we didn't evaluate effect of chemotherapy.

Although CT shows calcifications in neurocytoma but of great amount of blood vessels and drop metastasis, it is seen better on MRI. In two mentioned situations chemotherapy approach is benefit.

The differential diagnosis should be considering with other CNS tumors such as intra ventricular oligodendroma, astrocytoma and ependymoma (absence of cysts and calcifications).

Differential diagnosis of central neurocytoma in imaging depends on involved site and patient's age. Tumors involve lateral ventricle in young adults include oligodendrogliomas, subependymal giant cell astrocytomas, ependymomas, and low grade or pilocytic astrocytomas. Astrocytomas and ependymomas may involve other sites of ventricular system, but usually without intratumoral cysts and calcifications. Intratumoral calcifications in oligodendrogliomas are typically large and irregular [30].

Conclusion

Central neurocytomas with aggressive or malignant behavior, although rare especially in pediatric age group, have poor clinical outcomes. Treatment of choice is surgery and RT and chemotherapy have an accessory role in cure and long term survival depend on vast of surgical resection.

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