Case Report

Supratentorial Embryonal Tumor in Adult Patient: Case Report and Literature Review

Abstract

Embryonal tumors are the new nomenclature of the primitive neuroectodermal tumors or PNET. Their supratentorial location and their manifestation in adult population are not very frequent. Embryonal tumors are conformed from undifferentiated neuroepithelial cells that have the ability to show differentiation to several cell lines. Next is presented a case from an adult male patient with a clinical headache pictures and convulsions. With imaging study that shows a meningeal enhancement with frontal lobe infiltration with histopathological study of embryonic tumor with areas of glial differentiation.

Keywords: *Adults, embryonal tumors, supratentorial*

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Introduction

Embryonal tumors are the new nomenclature of the primitive neuroectodermal tumors (PNETs) and other central nervous system tumors according to 2016 World Health Organization (WHO) classification.[1] The classification of nonmedulloblastoma teratoid rhabdoid teratoid embryonic tumors has been a challenge. Previously called supratentorial primitive neuroectodermic tumors (PNET), associated with a poor neurological prognosis.[1]

The annual incidence in North American and European population is of 3/100.000 inhabitants under 15 years old. These have two locations: supratentorial and infratentorial, the infrantentorial location is more frequent where medulloblastomas are 20%–25% of all pediatric brain tumors. These tumors are malignant with high cellularity predominant in the pediatric population and very rare in the adults.[2]

Next, we will describe a supratentorial embryonal tumor case in an adult patient with its imaging characteristics and patient management.

Case Report

A male patient, 55 years of age, consulted for a 3-month picture of headache of the left frontal predominance, stabbing type, intensity 8/10 according to the visual analog scale of pain, persistent, which is exacerbated with Valsalva maneuvers, associated with nausea and emesis occasionally. It has been disabling during the last week, also associated with horizontal diplopia with a false image on the left, which improves with the occlusion of one eye and moderate photophobia. It denies improvement with common analgesics and interrupts the sleep pattern.

The neurological examination shows convergent strabismus of the left eye with limitation for abduction of the left eye, as the only positive finding to the physical examination, so it was decided to perform lumbar puncture with finding of opening pressure in 21 cm H2O after which patient presented headache improvement; a cytochemical study of CSF is performed discarding tuberculous meningitis and neurosyphilis, cytologic of CSF discarding neoplastic process and autoimmune process discarded.

Subsequently, a week after, the patient presented neurological deterioration due to increased intensity of headache associated with dysarthria, compromise of the left hypoglossal nerve, so that cerebral nuclear magnetic resonance imaging (MRI) was performed for study [Figure 1].

How to cite this article: Muñoz Montoya JE, Rodríguez PS, Duarte Mora DP, Benavidez Rojas MS, Cortes Jaramillo EJ, Chacón Zambrano LA. Supratentorial embryonal tumor in adult patient: Case report and literature review. Asian J Neurosurg 2020;15:733-6.

Submitted: 16-Jan-2020 **Revised:** 08-Feb-2020
Accepted: 31-Mar-2020 **Published:** 28-Aug-2020

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Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_19_20

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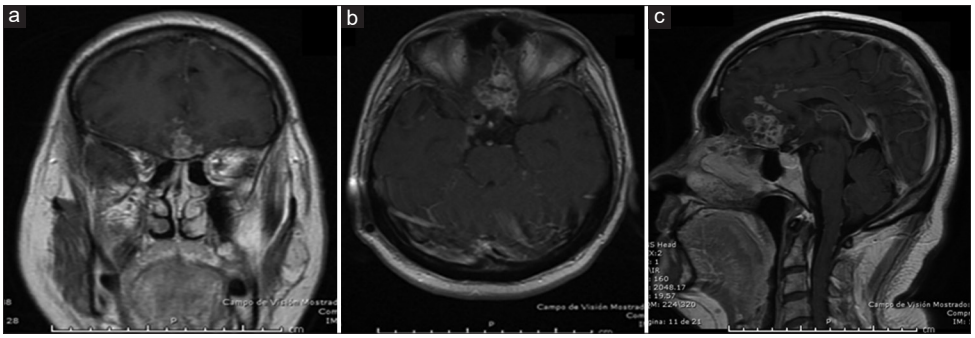

Figure 1: Nuclear magnetic resonance with contrast: (a) Coronal section where there is enhancement in the left front midline; (b) Axial section where baseline frontal enhancement is identified in the midline; (c) Sagittal section where basal frontal meningeal heterogeneous enhancement that infiltrates the frontal lobe and perilesional vasogenic edema is identified

It was decided to take the patient to transcribriform biopsy finding a slightly thickened and vascularized dura, and due to the persistence of endocranial hypertension a ventriculoperitoneal shunt is performed, a new MRI of the brain is performed [Figure 2], with a decrease in frontal lesion, then in the histopathological study, it was obtained as a report of embryonic tumor pathology with areas of glial differentiation [Figures 3 and 4], so it starts chemotherapy with etoposide, cisplatin, and cyclophosphamide for 6 cycles at the end of which radiotherapy and vincristine were added, evidencing a decrease in the size of the injury. In the follow-up of the patient at 16 weeks, the patient re-enters the hospital for convulsive episodes, then in said hospitalization it dies due to convulsive epileptic status.

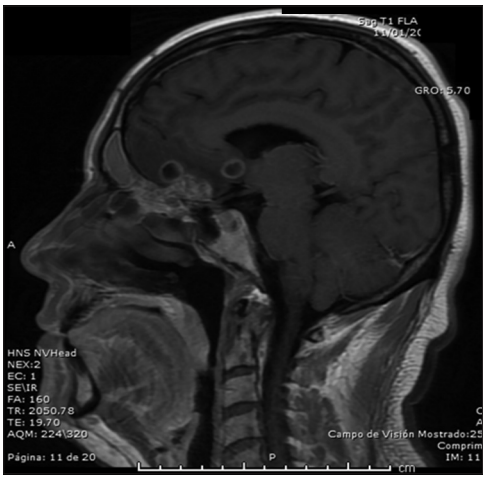

Figure 2: Contrasted nuclear magnetic resonance: Sagittal section where decrease in basal frontal lesion with vasogenic edema is identified

Discussion

Embryonic tumors or primitive neuroectodermal tumors (PNET) are tumors a heterogeneous group of malignant tumors, which can affect the central and peripheral nervous system.[3,4] Central embryonic tumors are rare neoplasms that may have a supratentorial, infratentorial, brainstem, or spinal cord location; they are more frequent in the pediatric population and are extremely rare in the adult population (over 20 years old). They represent 2.5% of cranial tumors in children and 0.46% in adults. Peripheral neuroectodermic tumors are derived outside the central and autonomic nervous system.[4,5,6]

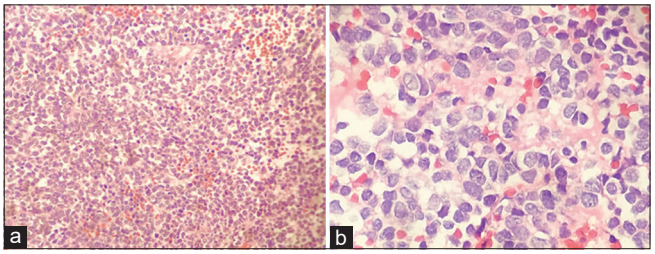

Figure 3: (a) Magnification shows proliferation of embryonic-looking tumor cells; (b) Increased increase are identified cells with small nuclei, prominent nucleoli and dispersed chromatin, the cytoplasm is clear and scarce

Embryonic tumors are an entity that rarely occurs in adults. It is defined as an embryonic tumor formed by undifferentiated neuroepithelial cells that have the ability to show divergent differentiation along neuronal, astrocyte, ependymal, muscular, or melanocytic lines. They are a very poor prognosis entity that, together with other factors such as tumor dissemination, tumor necrosis, metastasis, not receiving radiotherapy, and not achieving total resection, make it a fatal neoplasm.[7,8]

As in our case, where it was diagnosed in a young adult male patient, located in the frontal region with a report of embryonic tumor with areas of glial differentiation, with poor prognosis criteria since its resection was very difficult due to its tumor location.

Macroscopically, embryonic tumors are lobed, grayish or pinkish purpuric masses. Histologically they are small uniform cells, rounded or oval, not differentiated with little cytoplasm, hyperchromatic round nuclei, with high mitotic activity. Microscopically, they present calcifications, necrosis and "Homer-Wright rosettes."[3,4,9]

Immunohistochemical studies have demonstrated the expression of MIC2 and CD99 glycoprotein. Tumors with the MIC2 gene (CD99) appear to be less aggressive

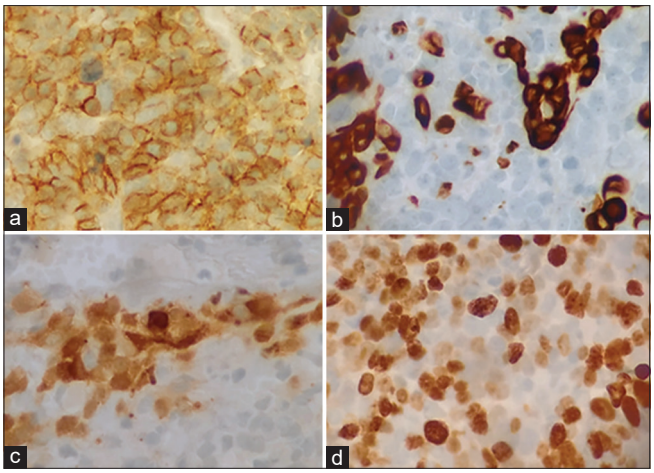

Figure 4: Immunohistochemical studies presented: (a) CD56 with membrane reactivity, which favors neural origin; (b) Enolase with focal reactivity, marker related to tumors of neuroectodermal origin; (c) PAFG with focal reactivity, which implies glial differentiation of tumor cells; (d) Ki-67 cell proliferation rate reaches 80% in tumor cells

after complete resection.[3,5] The molecular subgroups of embryonic tumors have a differential expression of the LIN28 and OLIG2 cell lineage markers. Tumors that have more overexpression of LIN28 are associated with greater aggression while tumors with low expression of LIN28 and OLIG2 were associated with a higher incidence of metastasis.[8,10]

Another molecular marker is Rad 51 which has an important role in the repair of homologous recombination by facilitating the transfer of chains between the broken sequences and their undamaged counterparts. Overexpression of this gene has been observed in most human tumor cells. Rads 51 is not only involved in carcinogenesis, but it is also associated with the affectation of radiotherapy resistance, also the overexpression of Rad 51 is related to poor clinical prognosis.[8,11,12]

The clinical manifestations of embryonic tumors in the adult population are presented according to the anatomical location of the tumor. Clinically, they may present with increased intracranial pressure, headache, nausea, emesis, confusion, seizures, and focal neurological deficits such as paresis, aphasia, facial paralysis, visual field defects, and papilledema due to rapid tumor growth.[3,13,14]

These clinical manifestations described previously, were identified in our described patient, associated with this, he presented endocranial hypertension, so he required management with peritoneal ventricular bypass valve.

For its diagnosis, the images were very useful not only for diagnosis but for surgery planning and planning of the craniospinal radiotherapy or intrathecal chemotherapy,[9] the cerebral MRI being the image of choice because it provides a better anatomical definition of the tumor. A diffusely disseminated neoplasm can be observed, on other occasions a well-defined solid mass, heterogeneous with cystic areas and necrosis can be evidenced. Its solid parts are hypointense in T1 and hyperintense in T2. In other cases, it may occur with intratumoral hemorrhage and focal calcifications. In adults, supratentorial embryonic tumors may present with moderate to severe surrounding edema.[3,4,6,9,13,15] Angiography can show focal areas of prominent vascularization within the tumor and correlate with the areas of nodular contrast.[9]

As for the standard treatment for supratentorial primitive neuroectodermal tumors whenever possible, a complete surgical resection should be performed followed by chemotherapy and radiotherapy. Multimodal treatment improves the results for patients with supratentorial embryonic tumors; however, in some cases, surgical management is not feasible due to the wide multifocal dissemination. Survival rates for adults are worse than children.[6,8] The 5-year survival of CNS embryonic tumors remains less than 35% in all age groups.[3,4]

Conclusion

Supratentorial embryonic tumors in the adult population are a rare entity, with nonspecific neurological manifestations that depend on their location, have wide focal dissemination, so that complete surgical resection is difficult, with a low survival rate despite multimodal management which has a reserved prognosis and high mortality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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