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

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 cytotoxic study of CSF is performed discarding tuberculosis 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].

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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.

**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] 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.
The molecular subgroups of embryonal tumors may present with moderate to severe surrounding edema. Angiography can show focal areas of prominent vascularization within the tumor and correlate with the areas of nodular contrast.

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. The 5-year survival of CNS embryonic tumors remains less than 35% in all age groups.

**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.

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**Conflicts of interest**

There are no conflicts of interest.

**References**

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