Occipital and Parietal Central Nervous System Embryonal Tumor NOS in an Adult Patient : A Case Report

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Introduction:

CNS embryonal tumors are highly malignant undifferentiated or poorly differentiated tumors of neuroepithelial origin, they take origin from embryonic cells that remain in the brain after birth, These tumors tend to spread through the cerebrospinal fluid (CSF) to other parts of the brain and spinal cord.Central nervous system embryonal tumor, not otherwise specified (NOS) was previously called primitive neuroectodermal tumors (PNET), these are rare tumors that form in the brain and spinal cord. (1)

We describe CNS (NOS) that arose in the occipital lobe of a 27 year old patient.

Case report:

This 27-year-old right-handed patient first presented with a homonymous hemianopsia and signs of increased intracranial pressure. The CT of the brain scan showed the right intracranial parietal and occipital expansive tumor responsible for an internal falcorial and temporal herniation. Due to clinical signs and symptoms consistent with raised intracranial pressure, she underwent urgent maximal debulking surgery, with post-operative scans demonstrating near-total resection.

Histopathological examination of the resection specimen showed dense tumor proliferation consisting of layers of cells with a high nucleo-cytoplasmic ratio and little cytoplasm. Mitotic figures were easily identified, including necrosis (figure 1).

Cells show immunohistological staining for synaptophysin and CD56. (Figure 2)

Staining for cytokeratin, TTF-1, INMSI, neurofilament, GATA3, SSTR2A, and chromogranin-A was all negative. Glial markers (Olig2 and GFAP) were also negative, and the protein INI-1 was positive in approximately 60% of tumor cells. The histological findings were considered to be most consistent with CNS-PNET, WHO grade IV, now known as a CNS embryonal tumor (NOS).

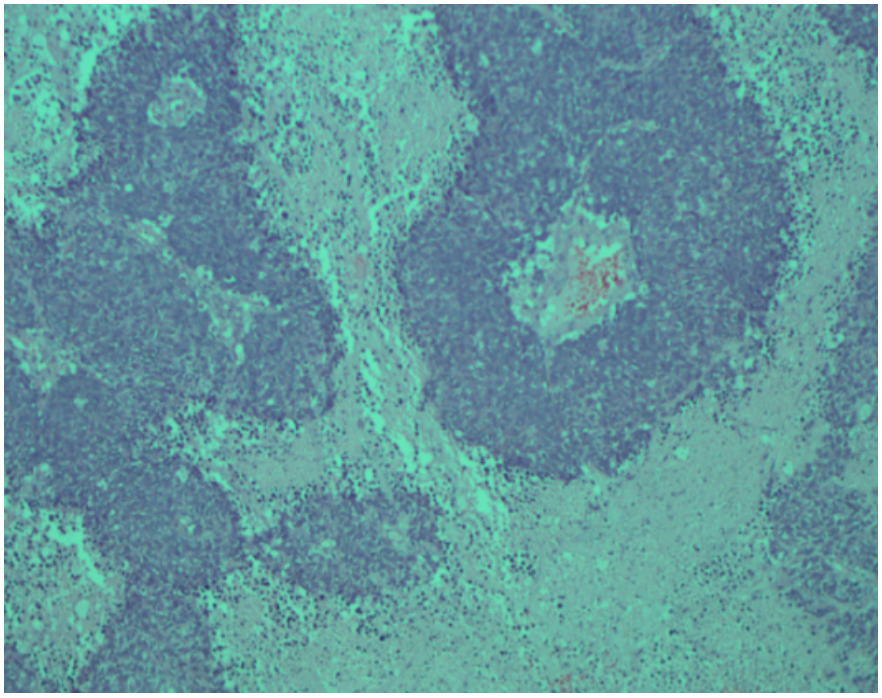

Figure 1 :CNS Emryonal tumor. HEx200. Largely necrotic tumor proliferation. Tumor cells have high nucleo-cytoplasmic ratio and show multiple mitotic figures.

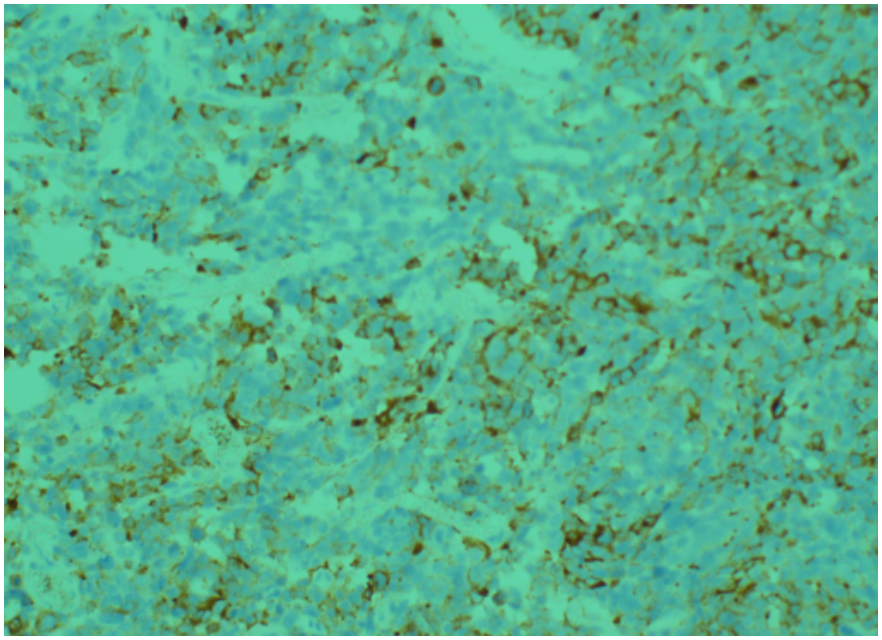

Figure 2 : CNS Emryonal tumor. IHC. Synaptphysin. Tumor celles express neuronal marquer. HEx20

Our patient underwent a thoracic, abdominal, and pelvic CT scan that showed pulmonary embolism with no secondary localizations of the tumor, a cerebral residual tumor of 4 cm, and a bone scan that was normal.

Seeing the recurrence of the patient's symptoms after two months, an MRI was performed that showed tumor recurrence with edema and hemorrhagic areas and meningeal carcinomatosis. Another surgery was performed with total resection, with cerebrospinal fluid analysis revealing no malignant cells. A second recurrence of intracranial hypertension symptoms led to the discovery of areas of meningeal enhancement involving the supratentorial region of the brain.

The patient began to receive two of the planned eight cycles of platinum- and alkylator-based chemotherapy. This initially consisted of cisplatin, vincristine, and cyclophosphamide (VIP), with improvement in the patient's presenting symptoms.

Discussion :

According to the 2016 WHO CNS classification, CNS embryonal tumors (NOS) are embryonal tumors other than medulloblastoma, atypical teratoid or rhabdoid tumors, embryonal tumors with multilayered rosettes, medulloepithelioma, CNS neuroblastoma, and CNS ganglioneuroblastoma.(2)

CNS (NOS) is a category for the tumors previously called PNET, which cannot be classified under a genetic or molecular group. With the "NOS" designation (not otherwise specified), there is insufficient information for classification. Some of these tumors were previously named embryonal tumors with abundant neuropil and true rosettes. (3)

They are still considered a nosological entity with distinct biological behavior; they account for less than 0.5% of all brain tumors and predominantly affect children aged 0–14 (2). The incidence of adult CNS embryonal tumors is difficult to determine because of their rarity and lack of signature biomarkers.

In supratentorial locations, vomiting, seizures, and headaches are common. Hemiparesis is present if the tumor affects the cortical motor areas or the descending tracts. The presentation is also affected by age. Younger patients present with irritability, vomiting, and visual problems.(4)

In our case, our patient had symptoms of supratentorial presentations that included raised intracranial pressure and a focal neurologic deficit.

In most cases, ETNOS are hypercellular lesions composed of poorly differentiated cells exhibiting round to oval crowded nuclei with stippled chromatin, a high mitotic index, and frequent apoptotic bodies. Overall, neoplastic cells are positive for synaptophysin, GFAP, INI1, CD99, and vimentin and may also express NFP, EMA, p53, and CKM.(2)

The treatment consists of complete surgical excision; chemotherapy and radiation are necessary adjuncts. Generally, they have a poor prognosis. (5)

Chemotherapy varies with each protocol, but combining vincristine, cisplatinum, cyclophosphamide, and etoposide is common. Bevacizumab is used to block vascular endothelial growth factors. Intrathecal methotrexate and topotecan can be included in the treatment protocol.(4)

Conclusion:

CNS ETNOS are highly aggressive tumors that predominantly affect children and are extremely rare in adults. They are believed to arise from primitive neuroepithelial cells, and the lack of signature biomarkers and genetic/molecular characteristics makes their classification challenging.

Acknowledgment

None.

Conflict of interest

All authors declare that they have no conflicts of interest.

Author's contributions

Nour Ghammem: collected the patient data and drafted the manuscript.

Amina Mokrani : revised the manuscript and gave critical review of the content.

Nada Mansouri: provided the histopathology images along with their comments for the manuscript.

Amel Mezlini : gave approval for the final version to be published after review.

Ethical approval

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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