Giant posterior fossa mature teratoma with adjacent subacute haematoma, compressive on the brainstem, with acute hydrocephalus. Case report

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Abstract: Mature teratoma of the vermis is a rare entity in neurosurgical adulthood pathology. We present the case of a 65 years old patient, admited as an emergency for intense headache (VAS 8/10), nausea, vomiting, gait ataxia, orizontal nistagmus, dismetria, disdiadocokinezia, predominant on the left side, long tracts signs, predominant on the left side. Native and contrast CT and MRI scan of the head revealed a tumoral lesion, in the vermian, paravermian and in the fourth ventricle, with the aspect of a teratoma with intratumoral subacute haemorrhage including a giant lesion 5,5/5/4,5 cm, compressive on mesencephalon, and with suprajacent acute internal hidrocephalus. Emergency neurosurgery was performed (occipital infratentorial craniectomy, microneurosurgical total tumoral resection and haematoma evacuation). Postoperative, the patient recovered progressivelly , subtotal neo and arhicerebellar symptoms. The motor long tract signs recovered slower and persisted incomplete.

Abbreviations: Visual autologus pais scale - VAS, Scor Karnofsky- SK, Computer Tomograf- CT, Magnetic Resonance Imaging- MRI, Cerebrospinal Liquid-LCR, CEA-Carcino Embrionar Antigen

Key words: Giant tumor, Posterior fossa, mature teratoma

Introduction

Introduction: Mature teratomas are germ cell tumors differentiated into all three germ layers with very low incidence (0,2%)1. Posterior fossae teratoma is a lesion with high morbidity (by neo and arhicerebellar syndrome) and mortality (by direct compression on mesencephalon and/or pons, by compression on the fourth ventricle with acute internal hydrocephalus and acute intracranial hypertension). We present the case of a patient admitted in our clinic for a posterior fossa and fourth ventricular tumor with intratumoral haemorrage compressive on

mesencephalon and acute internal hydrocephalus. The surgical intervention is urgent and mandatory because of the severity of this tumor and in this particular area.

Case Report: A 65 years old patient was admitted in our hospital as an emergency, presenting headache for a few months, suddenly increased within the previous seven days, which became intense (VAS 8/10), drowsiness, frequent vomiting, gait ataxia with large base, left 6th nerve incomplete paralisya, left disdiadocokinesia, left long tact signs, predominant motor, SK 70. CT scan of the head revealed a giant posterior fossa 5,5/5/4,5 cm tumor, vermian and left paravermian, with gadolinofil mural nodule 21/23 mm with calcified nodules, with hyperdense lesion adjacent with significance of subacute intratumoral haemorhage compressive on mesencephalon, with supraiacent internal hydrocehalus, suggestive for a teratoma with subacute intumoral haemorhage.

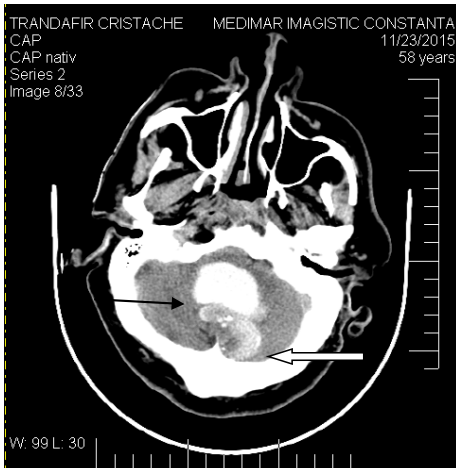

Figure 1 - Preoperative aspect: Cranial CT Scan with contrast substance: Vermian posterior, left paravermian and in the fourth ventricle tumoral lesion, moderate heterogeneous with a hypodense part having 3 calcified microopacities (white arrow). Anterior vermian and left paravermian haemorrhage (black arrow). The 2 lesions together were compressive on the mesencephalon and on the adiacent cerebellar hemispheres

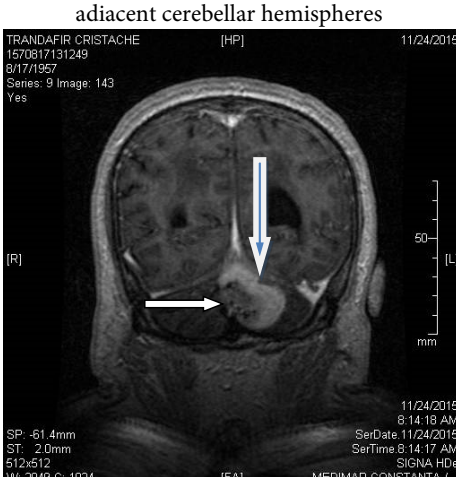

Figure 2 - Preoperative aspect. Cerebral MRI, T1 with Gadolinium coronal incidence: Central vermian and left paravermian heterogeneous lesion, with a posterior and central part of about 2,2/1,5 cm hypointensity, having inside small hyperintense areas (white arrow). Adiacent, superior vermian and left paravermian subacute haematoma (white and blue arrow)

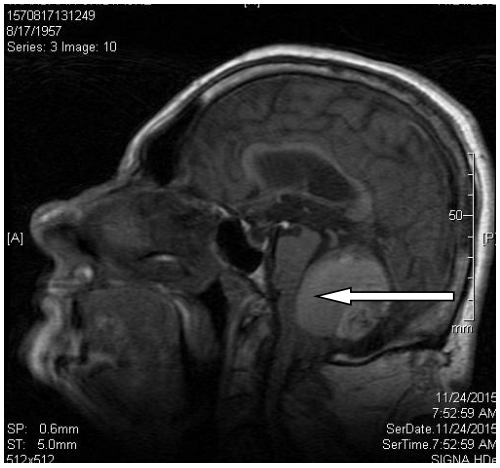

Figure 3 - Preoperative aspect. Cerebral MRI, T1 with Gadolinium enhancement sagital view: giant tumoral and hemorrhagic lesion described in figure 2 in the vermis and the fourth ventricle (5,5/5/4,5 cm), severelly compressive on the ponto-mesencephalic junction with supraiacent acute internal hydrocephalus (white arrow)

Ophtalmological examination revealed bilateral papilarry edema. Corticosteroids were administered 24 hours preoperative, which diminished the drowsiness. Emergency neurosurgery was performed, consisting of occipital infratentorial craniectomy,total microsurgical tumoral and haematoma evacuation.

Postoperative native and contrast cranial CT Scan revealed total resection of the tumor and total evacuation of the haematoma, with subsequent slight reduction of the internal hydrocephalus.

Postoperative, the patient developed chemical meningitis which was treated with corticotherapy and multiple serial lumbar punctures. After 2 weeks the chemical meningitis ceased, and the patient underwent progressive recovery of the arhi and neocerbellar symptoms. Long tract signs and the left sixth nerve paresis recovered slowly. Papilarry edema disapeared also. The patient was clinically, radiologically and ophtalmologically followed within the following 11 months.

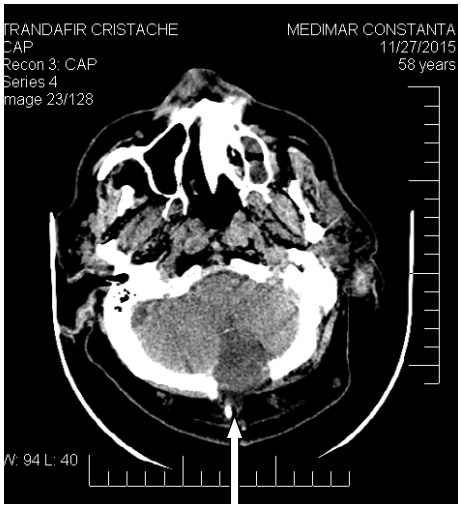

Figure 4 - Postoperative Control. Cranial CT scan with contrast: Total resection of the tumor and evacuation of the haematoma (white arrow)

Anatomopatologic exam revealed:

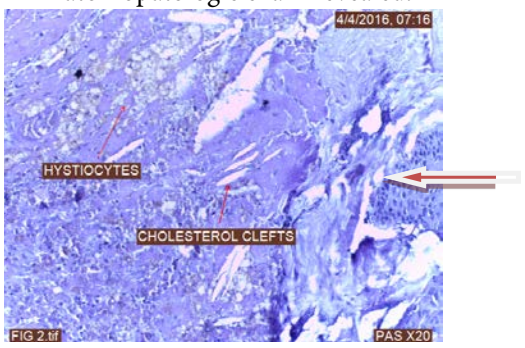

Figure 5 - Squamous stratified epithelium (red-white arrow) and the content of the clusters (cholesterol, hystiocytes-red arrows)

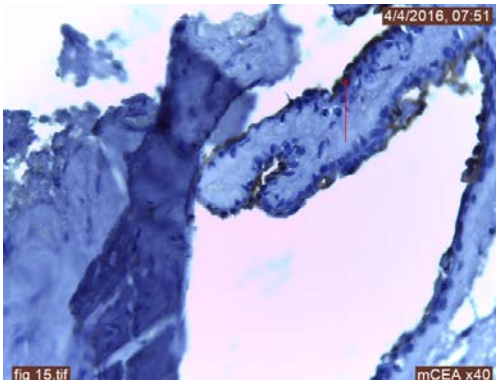

Figure 6 - Unistratified cubic epithelium which is lining the internal surface of the cystic tumor confirming the endodermal origin of the teratoma (red arrow)

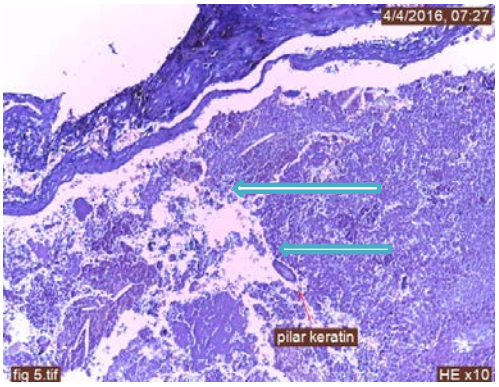

Figure 7 - Pilar keratin in the content of the cyst(red arrow). The ectodermal origin of the teratoma cyst was confirmed (blue–white arrows)

Discussion

According to Sanyal (1), teratomas are nonseminomatous germ cell tumors differentiating into all three germ layers. Intracranial mature teratomas are tumors with a very low incidence (0.2%), having a clear male predominance (5: 1). A posterior fossa teratoma is a rare occurrence (2, 5, 6).

According to Coulibaly (3), intracranial teratomas are congenital neoplasms most diagnosed in pediatric hood. Teratomas which involve the posterior fossa represent less than 0,5% of all intracranial tumors3. According to Bohara (4), standard histopathological examination for a mature teratoma showed a tumor with components of all the three germ layers. The histopathological differentiation between teratoma and dermoid cyst is very valuable for ruling out the presence of immature/malignant or germinomatous components that would require further adjuvant therapies (4). According to Desai and Goel (6), the outcome of teratoma is very poor6. According to Kondziolka (7) the mechanism which explains the hemorrhage into the tumor are the pathological deficits within the walls of the tumoral blood vessels such as hyalinization, necrosis or degeneration.

In the above described patient certain intratumoral haemorhage aggravated the evolution of the tumor by direct compression on the brain stem, acute internal hydrocephalus and increased intracranial pressure. Emergency neurosurgery was performed (occipital infratentorial craniectomy, total microsurgical tumoral resection and haematoma evacuation). Ventricular drainage was not compulsory. Multiple drainage lumbar punctures performed for the treatment of postoperative chemical meningitis accomplished well the treatment of internal hydrocephalus. Papilar edema ceased progressive postoperative. Postoperative cranial CT revealed total microscopic resection of the tumor and hematoma evacuation. The patient was

clinically and radiologically followed for the next 11 months after surgery.

According to my personal research, this is the first description of a giant mature teratoma in posterior fossa and the fourth ventricule with intratumoral haemorrage in an adult patient in Romanian neurosurgical literature.

Conclusion

Giant mature teratoma of the posterior fossa and fourth ventricle is a rare tumor in adults with high morbidity and mortality. Emergency neurosurgery (Craniectomy, tumor resection and haematoma evacuation) was mandatory because of the vital risk and also for the reduction of morbidities induced by the presence of the tumor.

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