A Rare Case of Intracranial Non-Germinomatous Germ Cell Tumor in a 21-Year-Old Romanian Male

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Introduction
Extragonadal germ cell tumors are a rare entity that is more prevalent in infants and young children, with preference to midline structures. The category of intracranial germ cell tumors is divided into pure germ cell tumor (GCTs) versus non-germinomatous germ cell tumor (NGGCTs). They usually present in the second decade of life with a male preponderance.

Case Report
We present here a rare case of intracranial NGGCT in a 21-year-old Romanian male, who presented with complaints of emesis, ataxic gait, and diplopia. A computed tomography scan of the head in the emergency department revealed a pineal-suprasellar mass along with obstructive hydrocephalus and dilated lateral and third ventricles without any bleeding. MRI of the cerebral, thoracic, and lumbar spine showed no evidence of leptomeningeal metastasis. The patient had elevated serum markers of beta-hCG, and AFP, which pointed towards a diagnosis of non-germ cell tumor as in pure GCTs these markers are normal. To relieve the obstruction from mass effect patient had an endoscopic third ventriculostomy (EVT). However, after the procedure, he developed central diabetes insipidus as a complication with a triphasic response. Biopsy of the mass revealed atypical cells with granular architecture and atypical glands with positive immune histological markers for NGGCT. These findings supported the diagnosis of mixed germ cell tumor with yolk sac carcinoma and seminoma components. Patient’s transient central diabetes resolved with normalization in his urine output. He was eventually stabilized and returned to Romania for further management.

Discussion
This case represents an acute presentation with challenging complications in the management of location-sensitive intracranial germ cell tumor. The patient presented with the symptoms of pineal mass, including diplopia, ataxia, headaches, and vomiting all which are signs of increased intracranial pressure. We did not perform a lumbar puncture and in note in cases such as this lumbar puncture should be avoided in these patients due to risk of herniation. Serum alpha-fetoprotein and beta-hCG levels were elevated, pointing more towards the diagnosis of mixed germ cell tumor or immature teratoma. For diagnosis and staging, a histologic examination is needed to establish a definitive diagnosis of an intracranial GCT. Surgery to obtain specimen is mandatory for patients with normal CSF, serum alpha-fetoprotein, and beta-hCG as pure germ cell tumor or mature teratoma must be distinguished from other lesions since the therapeutic approaches are different. Surgical biopsies can yield small samples which can lead to inaccurate diagnosis depending on the area where the tissue was obtained from as mixed tumors and contain various components, and a small area biopsy may only include one part. As a result, when tissue diagnosis is not reliable treatment should be based upon the outcome associated with most malignant history and worse prognosis. In SIOP CNS GCT study, the magnitude of alpha-fetoprotein elevation was correlated with adverse prognostic indicator with AFP>1000U/L revealing a progression-free survival rate of 32% versus 76% in those with AFP <1000 U/L (13). Also, in patients with recurrent NGGCTs prognosis is poor. In summary, intracranial germ cell tumors are rare brain tumors that should be distinguished based on histology and tumor markers as they will help in the guidance of therapy. An initial evaluation with neuroimaging, tumor markers, cytology from CSF, and biopsy is a must to distinguish further treatment and prognosis.

References