Cerebral Germinoma as a cause of Hydrocephalous in a 5m Infant

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Abstract
Germinomas are malignant intracranial germ cell tumors, usually found in pineal region. These tumors are exquisitely sensitive to radiation. Whole ventricle radiation (RT) with a boost field to the primary tumor is the standard treatment for localized germinoma. A 5m old infant came to our department of Paediatrics as a case of Congenital hydrocephalous diagnosed to be Cerebral Germinoma.

Keywords: Congenital Hydrocephalous, Germinoma.

Case Study
A 5m old male infant came to our department with chief complain of vomiting since 15 days of life which increased since last 1m and progressive head enlargement since 1m and loss of weight for 1m without any history of fever, loose stool, contact with Tuberculosis or birth asphyxia. That was a 1st order born out of NVD without any history of rash or fever during antenatal period. Immunized as per age and BCG scar present. Exclusive breast feeding since birth with low socioeconomic status. There was no h/o hospitalization. Birt weight was 2.1 kg and at 5m weight was 3.1kg. Head circumference was 46.5cm. RBS was 129 at the time of hospitalization. CBC, RFT, LFT, Electrolytes normal. Fundoscopy revealed normal fundus. CT Scan brain revealed Hydrocephalous with Transependymal edema predominantly in Lateral ventricles with evidence of ill defined hypodense SOL in sellar region extending left capsuloganglionic region. Confirmed with MRI Brain most probably as a case of Germinoma with CSF spread. Serum and CSF level of HCG was 52mIU/mL and 13mIU/Ml respectively which is high and suggestive of CNS metastasis. Patient was given Acetazolamide tablet and sent for Radiotherapy.

Figure 1 CT scan of that patient
Discussion
Germinomas, the most frequent intracranial germ cell tumors (IGCTs), are rare tumors of children and young adults. These tumors arise almost exclusively from midline structures. The two most frequent sites are the pineal gland and the suprasellar regions\(^1\), but they can also arise in the basal ganglia, thalamus, cerebral hemisphere, and the cerebellum\(^2\). IGCTs are malignant neoplasms arising from remnants of primitive germ cells that have failed to migrate to the genital crest during embryonic life\(^3\).

Clinical presentation depends on the location and size of the tumor: pineal region tumors can determine increased intracranial pressure due to obstructive hydrocephalus with headache, vomiting, somnolence and visual abnormalities. Suprasellar location can cause endocrine problems due to hypothalamic /pituitary axis dysfunction with diabetes insipidus, isolated growth hormone deficiency, delayed sexual development or precocious puberty and hypopituitarism.

Histologically IGCTs are classified in two different types: pure germinomas (50-70%) and non germinomatous germ cell tumors which include embryonal carcinoma, Yolk sac tumor, choriocarcinoma, teratoma and mixed germ cell tumors.

Diagnosis includes radiological exams (CT and MRI), measurement of tumor markers in blood and histological assessment.

Pure germinomas are highly radiosensitive, whereas NGGCTs are less sensitive to radiotherapy and they have a worse prognosis.

Radiotherapy, with or without chemotherapy, is the gold standard for the treatment of pure germinomas; surgery is sometimes needed in case of obstructive hydrocephalus\(^4\).

In Infancy almost all hydrocephalous are of congenital aetiology. But some belong to birth asphyxia, meningitis, hemorrhage etc as a cause. However IGCT as a aetiology of hydrocephalous is a rare entity. This stimulated us to publish this case.

Conclusion
The IGCTS are very rare and require particular attention in the diagnosis of neurological and endocrinological symptoms. Radiation therapy is the cornerstone of treatment for localized intracranial germinoma.

References