GERMINOMA OF THE THALAMUS - A RARE CASE REPORT AND REVIEW OF LITERATURE REVIEW OF LITERATURE.

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Abstract :
Germinomas of extrapineal region are extremely uncommon. The clinico pathological features of germinoma in the left thalamus, in a 9 year old male are presented in this report. Post-operative radiotherapy and chemotherapy is advocated.

Keyword : Germinoma, Germ cell tumor, left thalamus.

INTRODUCTION:
Germinomas are a type of Germ cell tumors (GCT). Intracranial germinomas constitute about 4.8% of all brain tumors in pediatric age group. The usual anatomical locations are pineal, parapineal and suprasellar regions. However, GCT are also reported to occur in unusual anatomical locations within the central nervous system like basal ganglia, thalamus, fourth ventricle, brainstem and spinal cord. Germ cell tumors originating in the thalamus and basal ganglia are extremely rare and so far only 42 cases have been reported in the literature.

It needs to be emphasized that GCT occurring in these unusual locations pose considerable diagnostic challenges and could affect treatment modalities. In this report we describe the clinical, imaging and histopathological features of a case of germinoma arising from thalamus.

CASE REPORT:
A nine years old boy presented in January 2011 with complaints of headache, vomiting, weakness of right upper limb and lower limb, difficulty in swallowing, broad based gait for the past ten months in the neurosurgery department. He was evaluated with routine investigations, blood tests and a Computed Tomography (CT) scan of the brain. The CT scan brain showed multiloculated heterodense cystic lesion in left thalamic region with areas of calcification (Fig 1). A stereotactic biopsy was done by the neurosurgeon and the histological, cytological examination was positive for germinoma (Fig 2 & Fig 3). Subsequently he received Radiotherapy (RT) to the primary tumor site (Whole brain RT 30 Gy and Boost 6 Gy).
The post radiotherapy CT scan brain revealed a residual lesion with gliotic changes in left thalamic region (Fig 5). Initial serum and spinal fluid markers of AFP and HCG was not done in radiotherapy department but Serum HCG was done after the patient was referred to medical oncology department and the value was 218 mIU/ml. He was planned for chemotherapy with cisplatin, etoposide and Ifosfamide with mesna. After the first cycle the patient defaulted and it was found later that the patient died due to severe diarrhea and fever probably due to neutropenic sepsis.

DISCUSSION:

It is reported that germinomas arising from basal ganglia and thalamus account for 4–20% of all intracranial germinomas, following those from the pineal and sellar region [5,6]. Absolute male predominance is the feature of this disease [7-10].
Most cases presented with single tumor, multifocality is rare. The clinical manifestations include mild weakness and numbness of limbs, nausea, and vomiting. Our patient presented with right-sided hemiparesis. The cognitive retardation and speech disorder is associated with involvement of the thalamus and its fibers. The involuntary movement is associated with impairment of extrapyramidal system. The symptoms progress slowly in most cases (2–24 months), while acute deterioration is attributed to intra-tumoral bleeding. The tumor involving the thalamus does not cause endocrine disorder such as precocious puberty, diabetes insipidus or disorder of eye movement. Compared with the tumors in other locations, the tumor in thalamus develops to a large size with no or minimal space occupying effect. Whereas the tumor in the pineal region compress the midbrain aqueduct and cause hydrocephalus and diplopia, and tumors of sellar region cause severe insipidus. Magnetic Resonance Imaging (MRI) and CT scans play an important role in diagnosis, while the serum AFP and HCG are normal. There are small or no high signal in T2-weighted images in the peri-tumor area that is attributable to demyelination process by the tumors slow growth pattern of tumor. This is in contrast to the presence of severe edema caused by malignant tumors like glioma and lymphomas. However, it can be easily confused with cavernous malformation. Cystic formation and calcification is also common in germinoma. A cyst formation was seen in our patient at diagnosis. Soejima et al., reported 6 cases with cysts and while 5 showed calcification. Higano et al. reported five cases with cyst and calcification in all six cases. The intra-tumoral bleeding not only leads to rapid deterioration but also leads to misdiagnosis of vascular malformations. In addition, it may also contribute to cyst formation. The main differential diagnosis is from malignant tumors such as gliomas and lymphomas. Many malignant gliomas also accompany with cysts. Therefore, the clues mentioned above are very helpful. Although surgical resection is not the first line for clarified diagnosed germinoma, sometimes the diagnosis is difficult preoperatively. Additionally, Sano et al. found half of the germinomas contain other cell types insensitive to RT, which would result to recurrence. Therefore, the surgical resection can fulfill the removal of the tumor cells insensitive to RT and lower recurrence. The intra-operative frozen section test is essential and very helpful to lessen morbidity. The extent of resection should not be over the rim of the tumor so that the postoperative neurological deficits would be minor and most would recover soon. Huh et al. had not found severe RT related neurological deficits in large prospective randomised trials dening the optimal treatment strategy are not yet available. The combined application of chemotherapy and RT or (adjuvant) chemotherapy alone remains debated and has to be investigated in large prospective randomised trials. In the present case, chemotherapy was started due to presence of residual lesions after completion of RT. Some authors reported chemotherapy can lessen the dosage of RT, and it is a good
supplement to RT. Chemotherapy after RT or solely chemotherapy is both reported with success. However, the complications should be controlled during chemotherapy, such as the severe infection and anemia. In the present case, patient died due to severe diarrhea and fever probably due to neutropenic sepsis after chemotherapy. Although germinoma of large size has a high risk of metastasis and recurrence, no significant difference of metastasis and recurrence were found in different locations.\textsuperscript{9}

CONCLUSION:
Outcome of pure germinomas is significantly better following a biopsy and chemoradiation. Chemoradiation is the reigning paradigm for the treatment of germinomas. Use of tumor markers, improved imaging technologies and safer biopsy techniques has made the diagnosis of intracranial germ cell tumors relatively straightforward.

References:


