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Case Report

Primary extragonadal germinoma of the pineal gland: a case report

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ABSTRACT

Primary intracranial germ cell tumors are rare and account for only 1% with the majority being germinomas. It occurs predominantly in adolescent males between the age group of 10-25yrs. These tumours occur predominantly in the pineal, parapineal or suprasellar region. Clinical presentation includes headache, nausea, vomiting, papilloedema, lethargy and somnolescence. Diagnosis of germinoma is important as treatment protocol differs when compared to the other histological subtypes. Here we are documenting a case of germinoma in the pineal region.

Keywords: Germinoma, Pineal gland

INTRODUCTION

The pineal gland is centrally placed in the brain and concerned with secretion of melatonin. Pineal tumours are uncommon and a heterogenous group of primary central nervous system tumours which includes germ cell tumours (GCT) which accounts for more than Fifty percent (> 50 %) and Non-germ cell tumours.[1,2] The germ cell tumours include germinoma and teratoma. Non-germ cell tumours include pineocytoma, pineal parenchymal tumours of intermediate differentiation and pineoblastoma. Primary melanomas of the pineal region are rare. These tumors are hyperdense due to high cellularity on CT and are iso to hyperintense on MRI. Tissue diagnosis plays a pivotal role in the diagnosis of pineal germinomas. Intracranial germinomas have an excellent prognosis on account of their sensitivity to radiotherapy with the 5 year overall survival rate being more than 90%.

CASE REPORT

A 21 year old male presented with headache and blurring of vision for a period of one month. He was investigated and found to have a hyperdense tumour in the pineal region on MRI. Subtotal excision of the tumour was done and the sample sent to us. Gross examination showed multiple gray white fragments measuring 2.5x2.5 centimetres in aggregate. Histopathological examination revealed a tumour composed of nests of round to polygonal cells having abundant eosinophilic granular cytoplasm with round to oval nuclei and single conspicuous nucleoli separated by delicate fibrovascular septa infiltrated by lymphoid aggregates. Immunohistochemistry showed diffuse positivity with Inhibin. With these findings a diagnosis of pineal germinoma was arrived.

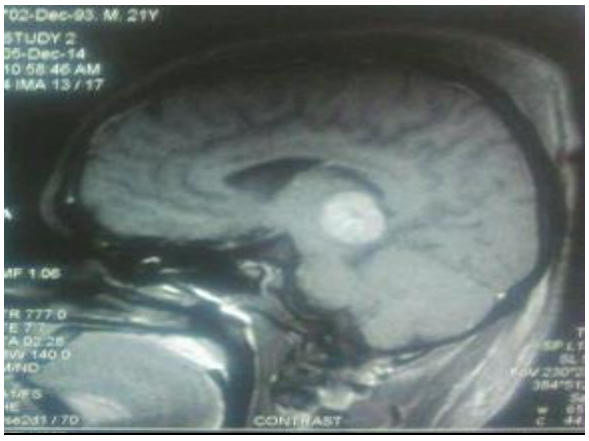

Figure 1: MRI picture showing a hyperdense lesion in the pineal region.

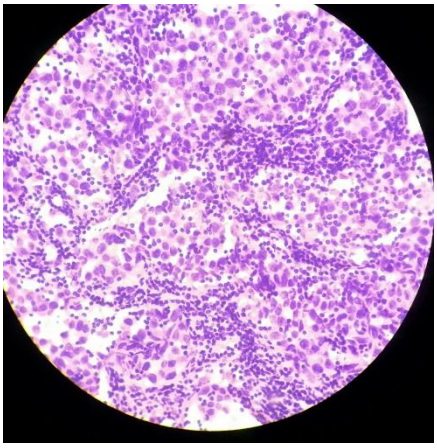

Figure 2: H& E, 40 x, Histopathology exhibiting nests of tumour cells separated by fibrous septa infiltrated by lymphoid aggregates.

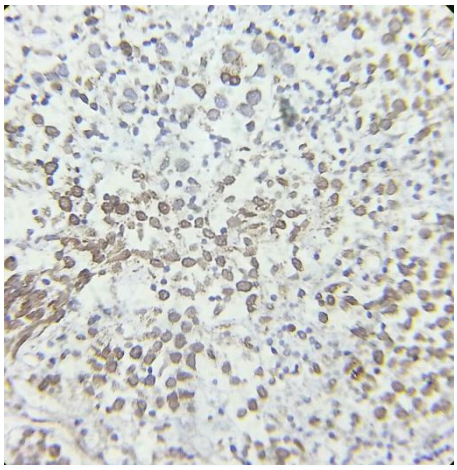

Figure 3: Immunuhistochemistry picture showing diffuse positivity in the tumour cells with Inhibin.

DISCUSSION

The term germinoma was first coined by Friedman.[3] Pure germinomas are the most common primary intracranial tumors accounting for 50% of all neoplasms in the pineal region. Less common locations for germinomas are intrasellar region, occipital region, fourth ventricle and cauda equina. 2-3% of all GCTs occur in extragonadal sites and Central nervous system is the commonest site of extragonadal GCTs. These tumors are predominantly midline lesions with occurrence more common in the pineal gland followed by suprasellar region, others being the thalamus and basal ganglia.Of the tumors of the pineal gland, germinomas account for 45%, NGGCT for 16%, pineal parenchymal tumors for 15%, gliomas for 17%, lipomas, cysts, metastases and meningiomas for 7%.

Cell of origin of CNS germ cell tumors according to the germ cell theory, are thought to be from the primodial germ cells which occur in the yolk sac during the third gestational week and migrate into the genital ridge in the sixth week, erroneously migrate to the pineal or suprasellar area where they undergo malignant change. According to the embryonic cell theory, germ cell tumors arise from mismigrational pluripotent embryonic cell.[4,5] Currently it has been postulated that these tumors arise from germinal elements at various stages of development.

Germinomas commonly occur in the first to second decade with median age of occurrence falling between 10-12 years. Men are more commonly affected than females with a sex predilection of 5:1 to 22:1. Chromosomal variations, variation in sex hormones and receptors are some of the proposed theories. According to the proposed theory of Sano, anterior neuropore closes earlier in males causing the enfolded embryonic cells to lie in the superficial part of the neural tube that subsequently forms the pineal gland.[5] Suprasellar region germinomas are more common in females.

Pineal germinomas are commonly seen in the midline and present with symptoms caused by increased intracranial pressure. Clinical presentation depends on the age of presentation, location and size of the tumor. Most common symptoms are headache, vomiting, endocrine abnormalities and visual changes. Most common presentation is the Perinaud syndrome which is seen in 34-50% of cases. Intracranial germinomas may also be associated with Downs syndrome[6] and Klinefleter syndrome.[7]

These tumours present with elevated levels of HCG and alpha-fetoprotein in the CSF and measurement of these tumor markers in serum and CSF aids in the diagnosis and follow-up of such tumours.

CT scan reveals a typical hyperdense lesion with central calcification and on MRI are iso to hypointense on T1 and T2 and show a vivid homogenous enhancement. Germinomas are prone to disseminate through the CSF and hence MRI of the entire neuroaxis and lumbar puncture are mandatory to rule out CSF seedling and drop metastases.[8,9]

Tissue biopsy is necessary for an accurate diagnosis due to the difference in treatment protocol for other histological subtypes. Germinomas are well circumscribed solid, soft and friable tumours with variable cystic component and calcification is a common feature. Currently World Health Organization classifies Central Nervous System GCT histologically as: Germino-Pure and with syncytiotrophoblasts, Non germinomatous germ cell tumour, mature and malignant teratoma, embryonal carcinoma, yolk sac tumour and choriocarcinoma. Tumors exhibiting more than one component are termed mixed germ cell tumours.

Histomorphology reveals lobules of uniform population of polygonal cells with distinct cell borders having pale to clear cytoplasm due to abundant glycogen and round centrally placed vesicular nuclei having a squared- off

appearance with single conspicuous nucleoli and separated by a delicate fibrovascular septa infiltrated by lymphocytes and activated macrophages. Lymphocytes in intracranial germinomas, thought to be reactive to antigens expressed by the tumour, are of T-lymphocytes in 70-80% cases and are of B-lymphocytes in 20-30% cases. Presence of macrophages and granulomas in intracranial germinomas as in testicular germ cell tumours supports the hypothesis of a common derivative. Differentials for germinoma are large cell lymphoma, metastatic carcinoma, melanoma and in granulomatous lesions, neurosarcoidosis.

Immunohistochemical marker for germinoma are placental alkaline phosphatase and CD 117. A more specific marker is the transcriptional regulator OCT 4. Serum and CSF PLAP analysis can be used as a useful marker for clinical follow-up and to monitor tumour relapses.

Germinomas are highly radiosensitive and chemosensitive. Radiosurgery is now considered as a treatment option along with chemotherapy and radiation. It has an excellent prognosis with the overall 5 year survival being 70-90% and 10 year survival being 70%. Genomic analysis has revealed that mRNA and miRNA serves as a novel therapeutic target.

CONCLUSION

Pineal germinomas have been rarely reported in literature and we herein present this case of pineal germinoma in a 17 year old boy with vague symptoms. Clinical work up with MRI, tumor markers, CSF analysis in conjunction with stereotactic biopsy and histopathological confirmation plays an important role in clinching the diagnosis of pineal germinomas.

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