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Intracranial Germinoma – A Case Report

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ABSTRACT

Intracranial germinomas belong to the class of germ cell tumors which are relatively rare intracranial tumors. Early recognition of this neoplasm is vital as germinomas are highly radiosensitive and effective/ early radiation therapy can result in relatively favourable overall prognosis. In this article we describe a 19 years old man who presented with pituitary tumor in the suprasellar region for which transsphenoidal decompression and biopsy was done. The histopathological examination confirmed it to be germinoma and he underwent craniospinal radiotherapy.

Keywords: Germinoma, germ cell tumor, radiotherapy

INTRODUCTION

Intracranial germ cell tumors are broadly classified into germinomas and nongerminatous tumors. Germinomas are being the commoner type with peak incidence in children and adolescents. Intracranial germinomas occur mainly in the pineal and suprasellar regions[1]. Less common sites include thalamus, basalganglia, ventricular system, cerebellum, frontal lobes and septum pellucidum. The clinical presentation depends upon the tumor location and size and it mainly includes endocrine abnormalities, visual field defects and signs of increased intracranial pressure[2]. The diagnosis of an intracranial germ cell tumor usually requires imaging and histological confirmation. Surgery is indicated to obtain a tissue specimen for diagnosis. Intracranial germinoma has relatively good prognosis when treated with radiation therapy [1, 2, 3].

CASE REPORT

A 19 years old man presented with progressive visual loss, recurrent episodes of head ache associated with vomiting over a period of several months. He also complained of generalized lethargy and loss of weight. He had no history of fever, seizures or trauma. On examination his vital signs were stable. He had bitemporal hemianopia with no other neurological deficits. His endocrine work up demonstrated panhypopituitarism and was subsequently placed on hydrocortisone, testosterone, thyroxine and desmopressin. MRI scan of brain revealed an enhancing lesion in the suprasellar region compressing the optic chiasma. An endoscopic transsphenoidal biopsy was done. Intraoperatively, erosion in the sella turcica floor was noted. Histopathology was consistent with germinoma. Serum tumor markers, alpha fetoprotein, beta human chorionic gonadotropin, cancer antigen, prostatic specific antigen and carcinoembryonic antigen all were normal. His ultrasound scan of the abdomen revealed no organomegaly. He was then referred to an oncologist and he underwent 28 cycles of craniospinal radiotherapy. During follow up his vision remained the same with no further deterioration of symptoms. He was lost to follow up after radiotherapy.

DISCUSSION

Primary intracranial germ cell tumors are rare central nervous system neoplasms. Germinomas are the most common accounting for 50-70 % of all germ cell tumors.[1] Most germ cell tumors occur before the second decade. Males are two times more likely than females to develop this tumor.[2] Approximately 95% of the primary germ cell neoplasm's are found in the midline, in the pineal and supra sellar region. The pineal region is the most common site for an intracranial germinoma compared to suprasellar region by a ratio of 2:1.[2, 3] Germ cell tumors will also occur in other sites including thalamus, basal ganglia region and fourth ventricle.

Clinical presentation of germ cell tumors depended on the location of the tumor in the central nervous system, the size of the lesion and age of the patient.[4, 5] Our case report highlights the unusual presentation in our patient, a 19 years old man with progressive visual deficits and symptoms of increased intracranial pressure confirmed to be having primary suprasellar germinoma. Suprasellar germinomas are observed more often in females and are the site

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of approximately 30% of intracranial germ cell tumors.

Germ cell tumors in the pineal region most commonly present with signs of increased intracranial pressure due to hydrocephalus, visual disturbances, pyramidal tract signs and ataxia. Symptoms such as nausea, vomiting and visual complaints can lead to an early diagnosis. The most common presenting symptoms for patients with suprasellar germ cell tumors are hypothalamic pituitary abnormalities such as diabetes insipidus, growth hormone deficiency and precocious puberty[2, 3]. Patients with isolated endocrine abnormalities are often diagnosed leter. Patients with suprasellar germinoma rarely present with signs of increased intracranial pressure[2]. Suprasellar germinomas are considered to have a poorer prognosis compared to those lesions arising in the pineal region[4]. Germinomas can disseminate the neuroaxis at any time in of course of the illness. The differential diagnosis of an intracranial germinoma includes craniopharyngioma, pituitary macro adenoma, metastatic cancer, malignant glioma, nongerminatous germ cell tumor and Langerhans cell histiocytosis[1, 3].

MRI with gadolinium is the diagnostic test of choice. It shows tumor size, vascularity, homogenecity and the relationship of the tumor to the surrounding structures.[5] Germ cell tumors are relatively isointense to normal white matter on T2 – weighted images and enhance with gadolinium. Calcification is likely to be seen in suprasellar and pineal region germinomas. CSF dissemination of primary intracranial germinoma throughout the ventricular system and subarachnoid space is common. Therefore before any surgical intervention, MRI of the entire spine is essential for the detection of metastasis[2].

The presence or absence of specific tumor markers produced and or secreted by tumor cells has been an extremely important adjuvant in the diagnosis of germ cell tumor.[4] Serum and cerebrospinal fluid tumor markers i.e. alpha fetoprotein and beta human chorionic gonadotropin may be helpful. A pure germinoma is usually not associated with elevation of these markers. The presence of elevated beta human chorionic gonadotropin [HCG] in patients with germinoma may correlate a less favourable prognosis.[6]

In the majority of clinical situations biopsy of the intracranial tumor is required for a specific diagnosis.[3] Endoscopic biopsy of the marker negative germ cell tumors is a safe and reliable method of establishing a diagnosis of germinoma. Intracranial germcell tumors encompass a variety of histological subtypes including germinomas and nongerminomatous tumors. Nongerminomatous tumors include highly malignant tumors such as embryonal carcinoma, choriocarcinoma, teratomas, which have extremely poor prognosis.[2, 5] The clinical manifestations, radiological characteristics and immunohistochemical evaluation of these tumors are similar to germinoma. Therefore accurate initial histological diagnosis and staging are very crucial to plan appropriate therapy since their treatment responses and prognosis differ considerably.[5] Germinomas are extremely radiosensitive, allowing the successful treatment without surgical debulking.[1, 2, 3, 4]. Thus surgery is required in the majority of patients for diagnosis alone[2, 4]. The role of extensive surgery is debatable. Radiation therapy has been the main modality of treatment for primary intra cranial germinoma.[1, 2, 3, 4] Whole brain irradiation may result in neurocognitive and endocrine sequelae. The rationale for the use of chemotherapy has been either to improve survival in these patients with disseminated disease or reduce sequelae, by limiting the amount of radiation in patients with localized disease[2]. Chemotherapy has been of greatest value in nongerminatous malignant germ cell tumor.

CONCLUSION

Germinomas are rare intracranial neoplasms which pose a challenge to clinician because of its variable clinical presentation with peak incidence in children and young adults .The diagnosis of these neoplasm requires imaging and histological confirmation. We describe this case in order to highlight the fact that early diagnosis of this rare locally destructive neoplasm is vital as these tumors are readily curable with radiation therapy alone or in combination with chemotherapy.

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