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A Rare Case of Primary CNS Germinoma in the Pineal Region with Long-standing Hearing Loss

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Abstract Germinomas in the brain are rare neoplasms which make up less than 1% of all intracranial tumors. Their clinical presentation is usually related to the increased intracranial pressure and/or hydrocephalus. This report describes a case who presented to our hospital for headaches and visual changes, including Parinaud's syndrome, that had started recently. The patient also had been complaining of chronic bilateral hearing loss for several years which seemed not properly evaluated previously. MRI brain showed a markedly enhancing lesion in the pineal region. CSF and serum tumor markers including B-human chorionic gonadotropin (B-HCG) and alfa-fetoprotein (AFP) were normal. Lumbar puncture with cytology was negative for malignancy. Biopsy was taken from the mass and sent to the pathology lab for testing. Histopathological examination showed changes consistent with germinoma and the immunohistochemical staining including that for placental alkaline phosphatase (PLAP) confirmed the diagnosis. There was no evidence of metastasis and the patient was treated with radiation-alone therapy that resulted in marked regression of the mass and significant clinical improvement except for the issue of hearing loss which had persisted, as expected, given its long-standing existance. In addition to highlighting the diagnostic evaluation and treatment approach for such rare tumors, this case suggests considering pineal germinomas among the differential diagnosis when dealing with hearing loss which needs careful evaluation and should never be underestimated.

Keywords: *CNS germinoma, pineal region, hearing loss, Parinaud's syndrome, immunohistochemistry*

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1. Introduction

Brain germinomas are rare neoplasms that represent around 1% of all intracranial tumors [1]. However, they are considered the most common type of germ cell tumor affecting the central nervous system. The most common site of involvement is the pineal region, followed by suprasellar region and other locations [2,3]. They usually occur in male children and young adults, with a median age of diagnosis between 10 and 12 years [3]. Here, we present a rare case of pineal region germinoma that was referred to our hospital for headaches and visual changes that had developed during the last few months. Moreover, the patient had chronic hearing loss for several years which is considered an unusual complaint for such tumors. When present, hearing problems secondary to germinoma can be reversed with debulking surgery if performed early enough before irreversible nerve damage occurs [7,8].

2. Description

18 year-old male was referred to our hospital for a 3-month history of headache that was left-sided and progressive in severity. It was associated with blurred vision and difficulty with ocular movements in both sides during the last few weeks. There was no nausea, vomiting, focal weakness or coordination problems. The patient had a past medical history remarkable for chronic hearing loss for several years that was not properly evaluated. He had no surgical history and his family history was unremarkable. He is a student in the high school for the deaf with an average performance. Physical examination revealed sensorineural hearing loss that was confirmed by audiogram. Moreover, he had some abnormalities consistent with Parinaud's syndrome including paralysis of upward gaze in addition to loss of accommodation and pupillary light reflex.

MRI of the brain with contrast showed a heterogeneous, markedly enhanced supratentorial central mass which is

expanding the left midbrain and extending to the left cerebral peduncle and the left thalamus (Figure 1). It was lobulated and irregular-shaped and measured 2.8 X 1.6 X 1.7 cm with mass effect on the adjacent third ventricle and ambient cistern. The lateral horns of the lateral ventricle were slightly dilated indicating early hydrocephalus. The patient was admitted and dexamethasone was started due to mass effect and cerebral edema in addition to levetiracetam for seizure prophylaxis. Laboratory investigations including B-HCG and AFP were unremarkable. Cerebrospinal fluid tests did not show significant findings and the cytology was negative for malignant cells. Therefore, a decision was made for neuronavigation-guided tissue biopsy for diagnostic purposes.

Histopathological examination revealed a mixed population of lymphocytes and large cells that have centrally placed nuclei, prominent nucleoli and abundant vacuolated cytoplasm (Figure 2). The tumor cells stained strongly for placental alkaline phosphatase (PLAP), but were negative for CD117, CD30 and alfa-fetoprotein (AFP). With these findings, the patient was diagnosed with pineal germinoma. MRI of the spine and computed tomography (CT) of the chest, abdomen and pelvis showed no evidence of metastasis.

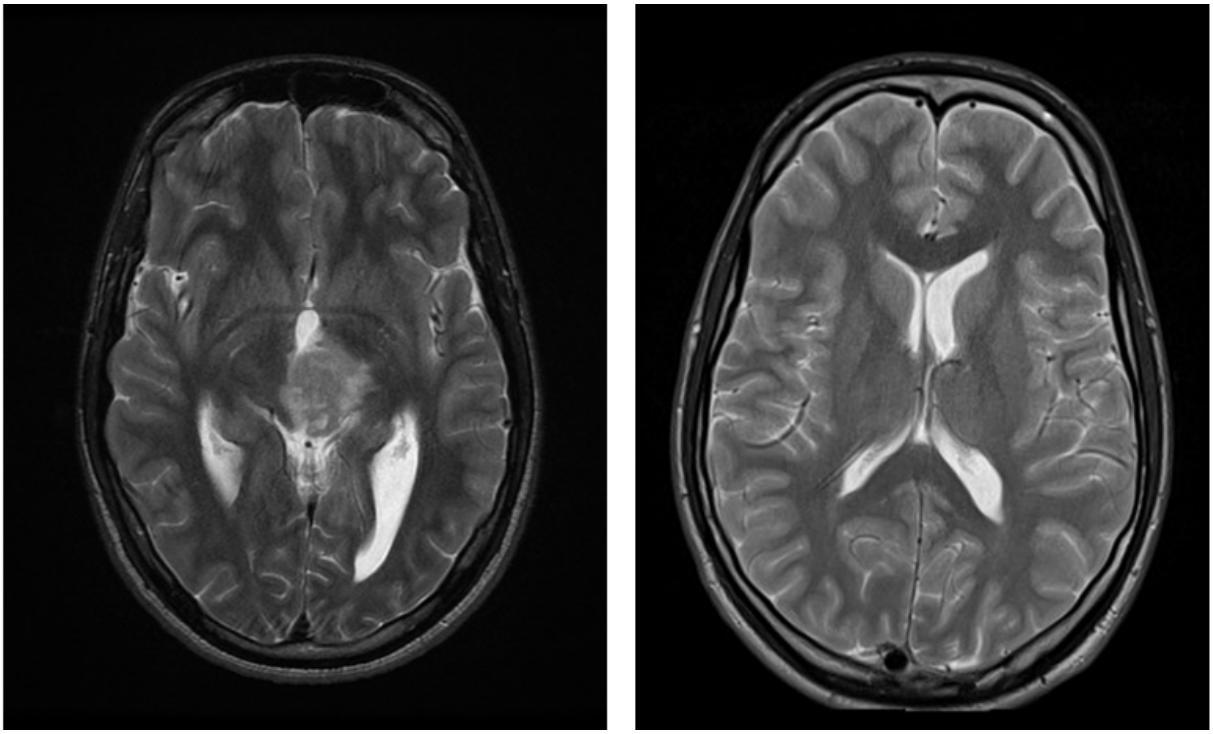

Figure 1. Brain MRI with contrast showed a heterogeneous, irregular-shaped central mass with marked enhancement in the pineal region (left). Repeat imaging following radiotherapy showed significant interval regression (right).

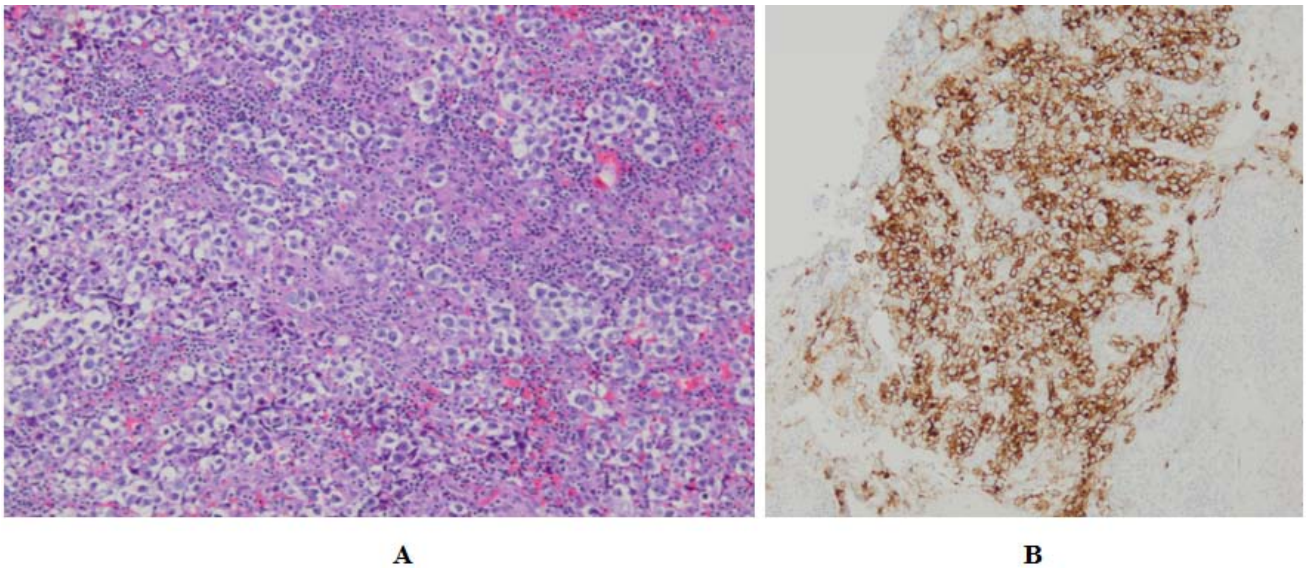

Figure 2. Histopathology of the brain lesion showed a mixed population of lymphocytes and large cells that have centrally placed nuclei, prominent nucleoli and abundant vacuolated cytoplasm(A). The tumor cells stained strongly for placental alkaline phosphatase (B).

The patient's hospital course was complicated by the development of some nausea and vomiting that was associated with gradually worsening mental status requiring intubation and transfer to the neurological intensive care unit. Repeat MRI brain revealed interval progression of the hydrocephalus, with a maximum bifrontal diameter reaching 4.1 cm. Neurosurgery was consulted and ventriculoperitoneal (VP) shunt was inserted with significant improvement in his conscious level. The patient was discharged home in a stable condition with a plan for outpatient radiation therapy that was based on a decision after a multidisciplinary meeting. Upon follow up after radiation, the tumor showed significant interval regression with marked improvement of mass effect leading to near resolution of his clinical manifestations except hearing loss which had persisted. Debulking surgery was not considered as it was felt unlikely to resolve the deafness issue given the chronic nature of the problem besides the excellent sensitivity of the tumor to radiation.

3. Discussion

Malignant germ cell tumors (GCTs) are usually found in the gonads (ovary and testis). Around 2% to 5% of these tumors can occur on the midline body structures and involve any part from the pineal region to the coccyx [4]. Central nervous system is considered the second most common location after the mediastinum for extragonadal GCTs, with the majority being pure germinomas [4,5]. In decreasing order of frequency, germinomas typically occur in the pineal recess (50%-65%), suprasellar region (25%-35%), and basal ganglia/thalamus (5%-10%). Although involvement of the pineal region, as seen in this case, by germinomatous GCTs is common, the tumors in this region are very rare and make up less than 1% of all intracranial neoplasms in children and young adults.

The clinical presentation of pineal germinomas is usually related to increased intracranial pressure and hydrocephalus and can include headaches, visual changes, confusion, nausea and vomiting. Neuro-ophthalmologic findings, classically Parinaud's syndrome, is a common presentation of such tumors due to the compressive effects on the dorsal midbrain [6]. All these manifestations were found in our patient, but he also showed hearing loss which is far less commonly reported in the literature. Moreover, it seemed to be the initial complaint in this case which had been going on for several years with no appropriate work up including brain imaging until late referral to our hospital. This symptom is explained by the proximity of the pineal region to the auditory pathways in midbrain, upper brainstem and diencephalon [7]. It is crucial to consider intracranial imaging when dealing with hearing loss and consider pineal germinoma among the differential diagnosis [7,8] even if the classical symptoms were lacking. Early surgical intervention can reverse hearing loss in some cases.

The best imaging modality for pineal germinoma is brain MRI with contrast which typically shows hyperintense mass with cystic and necrotic changes that markedly enhances following intravenous contrast administration [9,10]. MRI of the whole spine is also recommended in addition to lumbar puncture to evaluate for CSF seeding and drop metastases [3,9]. Measurement of tumor markers, such as B-human chorionic gonadotropin (B-HCG) and alfa-fetoprotein (AFP) can be helpful in the diagnosis and follow up of such tumors as germinomas only secrete B-HCG and the levels of AFP in the serum and CSF are usually normal [11].

There are some tumors that can mimic pineal germinomas and these include pineal parenchymal tumors, such as pilocytic astrocytomas and high grade gliomas, and nongerminomatous GCTs including teratomas and embryonal carcinomas [3]. Tissue biopsy is an important tool that should be done for reaching to a definitive diagnosis of pineal germinoma given the significant differences in treatment options compared with other histological subtypes found in this region. Surgical resection does not have a major role in the management of these highly vascular tumors [11]. However, they are very sensitive to radiation treatment and radiation-alone approach can provide cure with a five-year overall survival greater than 85% [11,12,13]. The main disadvantage associated with this approach is the increased risk of long-term complications associated with high radiation doses and volumes. For this reason, multimodality approach consisting of neoadjuvant chemotherapy followed by lower doses and volumes of radiation might be suggested to minimize radiation toxicity especially for metastatic disease and relapsed cases [11].

4. Conclusion

Pineal germinomas are very rare and represent less than 1% of intrancranial tumors. Similar cases have been rarely reported in the literature. The presence of hearing loss could be the only initial symptom which should alert the physician to the importance of considering pineal tumors in the differential diagnosis. Clinical work up usually requires MRI brain and spine, tumor markers measurement, lumbar puncture and tissue biopsy. The current standard of treatment is radiation treatment, with or without neoadjuvant chemotherapy, which provides long-term control of the disease.

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