# Case Report Metastasis germinomas of the external acoustic meatus: in a 19-year-boy: a case report

Rong Zhao1\*, Jiang Wang2\*, Ya Ba3, Qiong Zhang4

<sup>1</sup>Department of Nuclear Medicine, Urumqi General Hospital of Lanzhou Military Region, Urumqi, Xinjiang, 830002, China; <sup>2</sup>Department of General Surgery, Urumqi General Hospital of Lanzhou Military Region, Urumqi, Xinjiang, 830002, China; <sup>3</sup>Department of Nuclear Medicine, The First Affiliated Hospital of Xinjiang Medical College, Urumqi, Xinjiang, 830002, China; <sup>4</sup>Medical Department, Urumqi General Hospital of Lanzhou Military Region, Urumqi, Xinjiang, 830002, China. \*Equal contributors.

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**Abstract:** Background: The external acoustic meatus metastasis of germinomas is a rare event. Case presentation: we describe a 19-year-old boy with stuffiness and earplug of the left ear but no other symptoms. The whole body F-18 FDG PET/CT and the brain MRI are performed and demonstrated a germinoma in the pineal gland and vermis cerebellum region and in the right of thalamus. Photomicrograph showing marked infiltration of lymphoplasma cells and macrophages including giant cells. Immunohistochemical analysis results demonstrated the tumor cells are strongly positive for CD117 and PLAP. The final diagnosis was germinoma and all adjuvant therapy was achieved. Conclusion: Although the external acoustic meatus metastasis of germinomas is rare, the diagnosis should be taken into serious consideration in order to improve. In addition, F-18 FDG PET/CT was very useful in diagnosis primary disease and excluding distant metastases. To our knowledge, this is the first published report of this type of case.

Keywords: Germinomas, external acoustic meatus, immunohischemistry, F-18 FDG PET/CT

#### Introduction

Intracranial Germ cell tumors (GCTs) are relatively rare in Western series, accounting for 0.3-0.5% of patients with primary central nervous system (CNS) tumors. However, the occurrence of the tumors are far higher in Japan and in northeast Asia, where the reported rate about 3.0% of primary intracranial tumors [1-4]. Approximately 90% of Intracranial GCTs occur in the second decade of life. They most frequently occur in the pineal and suprasellar region, in general, pineal region germ cell tumors outnumber suprasellar tumors by a ratio of 2:1 [1, 5]. The tumors can also arise in other regions of the brain, with a tendency to develop in the midline, including in the fourth ventricle, basal ganglia and/or thalamus. In general, GCTs of the central nervous system which arise in the basal ganglia and/or thalamus are more likely to be germinomas, rather than other forms of germ cell tumors, and the histological subtype is the single most predictive factor of outcomes [6-16].

Many imaging characteristics of germinomas arising in these primary regions have been described, but the metastasis germinomas of the external acoustic meatus was rarely. In this report, we present a case of an atypical metastasis region of a germinoma in external acoustic meatus.

#### **Case presentation**

The patient was a 19-years-old boy. Medical history began 3 months before with the left ear stuffiness and earplug symptom. A consolidation area in the external acoustic meatus was found with temporal bone Routine CT (The information was lost). In order to obtain better treatment, the boy chose another hospital. On admission an obviously red neoplasm in the external acoustic meatus was observed during the conventional examination of otoscope (**Figure 1**). He had no remarkable medical or family history. There were also no other abnormalities. After the red neoplasm was completely removed, the above-motioned symptom of



Figure 1. An obviously red neoplasm in the external acoustic meatus is observed during the conventional examination of otoscope (arrow).



**Figure 2.** Histological features of the germinomas. Photomicrograph showing marked infiltration of lymphoplasma cells and macrophages including giant cells (H&E stain, A: 200x, B: 400x). Immunohistochemical analysis results demonstrated the tumor cells are strongly positive for PLAP (C: 200x) and CD117 (D: 200x).

the patient was immediately disappeared. Histological examination described a seminoma (Figure 2A, 2B). No tumor was found in some other sites, including mediastinum, testi-



**Figure 3.** The whole body F-18 FDG PET/CT was examined. The whole body F-18 FDG PET/CT scan reveal an intense FDG hypermetabolism in the pineal gland, vermis cerebellum region and in the right of thalamus on PET (upper row, arrow), as well as the portion was a slightly hyperdense than gray matter on CT (upper row, arrow). The surgery area of the external acoustic meatus show a slightly FDG hypermetabolism, and the corresponding parts show postoperative changes on CT (lower row, arrow).

cle. For further search for primary tumor, the whole body F-18 FDG PET/CT scan revealed an intense FDG hypermetabolism in the pineal, vermis cerebellum region and in the right of thalamus, as well as there are an hyperdense lesion in those regions (Figure 3 upper row). The surgery area of the external acoustic meatus showed a slightly FDG hypermetabolism, and the corresponding part showed postoperative changes on CT (Figure 3 lower row). The Brain magnetic resonance imaging (MRI) showed a lesion in the pineal, vermis cerebellum region and in the right of thalamus with signal intensity hypointense on T1-weighted images (Figure 4A), iso-intensity on T2-weighted images (Figure 4B) and hyperintense on FLAIR images (Figure 4C). The lesion was homogeneously enhanced with gadolinium-diethylene triamine pentaacetic acid (Figure 4D). Subsequently, Its immunohistochemical staining confirmed the diagnosis of germinoma (PLAP positive, CD117 positive) (Figure 2C, 2D). The patient received four courses of Chemotherapy. Adjuvant radiotherapy was then given. Treatment has now been complete for 11 months. At his most recent the brain MRI study showed no evidence of tumor recurrence.

#### Discussion

Germ cell tumors (GCTs) are classified into two categories: seminomas and nonseminomatous GCTs. It affects not only the gonads but also extragonadal tissue.

The testes and ovaries are the most common sites where GCTs occur. Seminomas are divided into two types, one type occurs in the gonad including testicle, prostate gland [17] and so on; The other type occurs outside of the gonad including mediastinum [18], central nervous system [19], retroperitoneal [20] and so on.

Seminomas, which are known as germinomas in the pineal gland and dysgerminomas in the



**Figure 4.** The brain was examined with MR imaging. A. Axial spin echo T1-weighted MR images reveal a homogeneously hypointense, space occupying lesion at the pineal, vermis cerebellum and in the right of thalamus (arrow). B. Axial spin echo T2-weighted MR images show iso-intensity lesion (arrow). C. The lesion is homogeneously hyperintense in the FLAIR images (arrow). D. Axial T1-weighted MR images show a homogeneous enhancement after an intravenous injection of contrast medium (arrow).

ovary, usually appear as large, bulky, solid masses with fibrovascular septa. A seminoma is not microscopically distinguishable from a gonads dysgerminoma or intracranial germinoma [21-26]. Just like in our report, since no more information, the tumor of the left external acoustic meatus was first considered for the seminoma by routine pathological results.

Germinomas are the most common tumor among germ cell tumors. It is not rare in Asian populations, with a male predominance. These malignant intracranial tumors usually occur in children and adolescents [10]. Their usual localizations are the midline central nervous system structures, most frequently the pineal gland. Our research has similarity with previous studies.

Intracranial germinomas may produce a series of classic neurologic syndrome, such as hemiparesis, precocious puberty, diabetes insipidus and so on [10, 11, 27]. However, there was no specific symptom in the early stage of this tumor. Tamaki N et al reported that the clinical course was extremely slow [10]. Only when the tumor pressured and affected the surrounding tissues and organs, symptoms appear. The vast majority of previous cases were diagnosed after dementia or hemiparesis had developed [7-12, 27, 28]. Local spread of intracranial germinoma within the brain and throughout the subarachnoid space [29], is not uncommon. This type of spread, moreover, is commonly limited to the soft tissues within the cranial cavity showing little or no prone to extracranial invasion. So, the external acoustic meatus metastasis of classic germinomas is extremely rare

except for a clinical case report of seminoma in the temporal bone [30]. To our knowledge, this is the first report that describes a metastasis germinoma in external acoustic meatus.

In our present case, the diagnosis of germinoma was difficult owing to the deficits of neurologic syndrome and more information. The whole body PET/CT imaging findings and the immunohistochemical studies are vital. First of all, a large number of tumor cell infiltrations were found in the neoplasm of the left external acoustic meatus. Biopsy confirmed the diagnosis of Seminoma. Secondly, for further search for primary tumor, the whole body PET/CT and the brain MRI were all showed that the tumor was located in brain. Just like Zimmerman RA et al had reported that both computed tomography (CT) and magnetic resonance imaging (MRI) are highly sensitive in the detection of pineal and suprasellar masses, as well as germ cell tumors in other regions of the brain [31, 32]. Thirdly, immunohistochemical studies confirmed further the diagnosis of germinoma. It was shown that the tumor cells were strongly positive for PLAP (placenta alkalinity phosphatase) and CD117. In the majority of clinical situations, biopsy of the germinomas is required for specific diagnosis. Immunohistochemical staining for PLAP and CD117 were well known [33, 34]. All results further proved that the tumor was originated from germinomas.

Because of germinomas localization and multicentricity, surgery rarely is curative. Germinomas respond readily to chemotherapy and radiotherapy, leading to a good prognosis and high rate of survival [35]. In our report, the patient had complete or near complete remission of the primary tumour, with a long survival outcome. However, if the diagnosis is delayed, the disease can become disseminated by local or CSF spread.

## Conclusion

This case illustrates an unusual metastasis localization of germinoma associated without more unusual tumour. Although early imaging and a biopsy-based diagnosis is important to determine early treatment and to help minimize any associated neurological deficits. We propose that primary intracranial germinoma should be considered in the diagnosis of head lesions in the second decade of life, especially those male patients without obvious signs of neurologic syndrome. Awareness of the possibility of external acoustic meatus involvement by germinomas is important because early radiation and/or chemotherapy can result in a cure of an otherwise locally destructive neoplasm. In conclusion, since metastasis germinomas of the external acoustic meatus is rare, the suggestive case described may help the clinicians to consider them in the differential diagnosis in order to improve the treatment.

Address correspondence to: Dr. Qiong Zhang, Urumqi General Hospital of Lanzhou Military Region, Urumqi, Xinjiang, 830002, China. E-mail: zhangqiong\_z@yahoo.com.cn

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