

Non-Metastatic Suprasellar Germinoma In A Child: Case Report

Çocukta Metastaz Yapmamış Suprasellar Germinoma Olgusu

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Özet

Hipotroidizm, diabet insipitus ve görme bozukluğu ile başvuran 11 yaşındaki kız olguyu sunduk. Manyetik Rezonans Görüntüleme (MRI)'de geniş suprasellar yerleşim göstererek optik kiazma ve hipofiz bezi sapına baskı yapan ve kontrast madde enjeksiyonu sonrası yoğun kontrast tutulumu gösteren lezyon saptandı. Tüm spinal kolonun kontrastlı MRI incelenmesinde patolojik kontrast tutulumu saptanmadı. Serum ve serebrospinal sıvıda alfa-fetoprotein (AFP) ve human koryonik gonadotropin (BHCG) gibi tümör markırları normaldi. Paranasal veya kemik invazyonu ve subaraknoid aralığa yayılım tespit edilmedi. Lezyonun total rezeksiyonu başarılı bir şekilde yapıldı. Histopatolojik incelemede germinoma tanısı kondu. Cerrahi sonrası, hasta kombine adjuvant kemoterapi ve radyoterapi ile tedavi edildi.

Anahtar Kelimeler: Germinoma, Manyetik Rezonans Görüntüleme, Tümör Metastaz

Abstract

Eleven-year-old girl presented with visual disturbance, diabetes insipidus and hypothyroidism. The lesion in magnetic resonance imaging (MRI) was established as a large suprasellar mass stretching the optic chiasm and pituitary stalk. The lesion showed intense contrast enhancement by intravenous administration of a contrast media. MRI examination of whole spine with contrast showed no pathology. Tumor markers such as alpha-fetoprotein (AFP) and human chorionic gonadotrophin (BHCG) in the serum and cerebrospinal fluid were at normal range. Paranasal or bone invasion and spread of subarachnoid space were not detected. Total resection of the lesion was successfully achieved. Histopathological examination revealed germinoma. After surgery, the patient was treated with the combined approach of adjuvant chemotherapy and radiotherapy.

Key words: Germinoma, Magnetic Resonance Imaging, Neoplasm Metastasis

Introduction

Germinomas comprise about 0.5–2.1% of all intracranial tumors and usually arise in the pineal or suprasellar regions (1). The pineal and suprasellar regions are the most common localization for intracranial germinomas although germinomas locate in other regions such as the thalamus and basal ganglia. They are much less frequent (2,3) within germinomas and account for only 4-10% of all intracranial germinomas (4,5).

Germinomas have a tendency to spread throughout the ventricular system and subarachnoid spaces. They have propensity to infiltrate adjacent soft tissue (6). We presented a case of primary suprasellar germinoma that compressed optic chiasm and slightly extended into cavernous sinus.

Case Report

In the current study, we reported the case of an eleven-year-old-girl who admitted to hospital due to irregularities affecting the field of vision, with clinical signs and symptoms such as polyuria, polydipsia and poor growth. There was short stature in her physical examination. Height and weight of the patient were less as compared to her age-matched children.

Diabetes insipidus was suspected because of the complaints of polyuria and polydipsia. MRI of the cerebrospinal axis was performed on 1.5 Tesla unit and MRI of the pituitary gland showed a large and solid mass within suprasellar region, extending into suprasellar cistern and hypothalamus superiorly and right cavernous sinuses.

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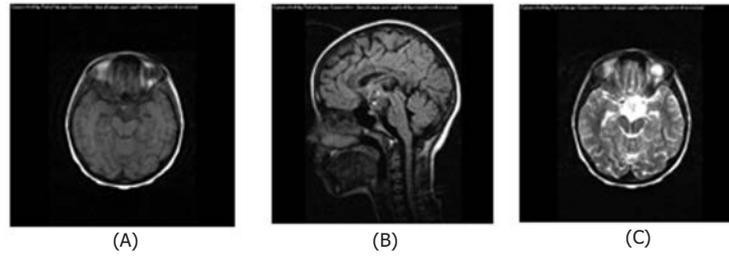


Figure 1: Transverse T1-weighted image (A) and midline sagittal T1-weighted image (B) showed an isointense, transverse T2-weighted image showed hiperintense in the suprasellar lesion (C). The lesion caused of optic chiasma and pituitary stalk. Intratumoral hemorrhage showed hiperintense focus in the mass both axial and midline sagittal on T1 weighted images.

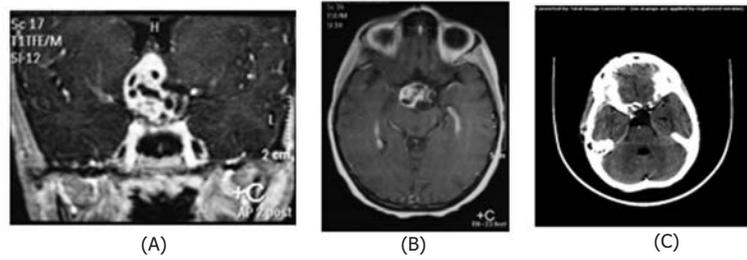


Figure 2: Intravenous administration of contrast media, the lesion showed intense contrast enhancement axial and coronal on T1 weighted images (A-B). There was no residual tumor in supracellar area on post operative axial CT image. Craniectomy defect and subdural hemorrhage was observed on right front-temporal region (C).

The optic chiasm and hypothalamus were compressed by the lesion. The lesion showed isointense signal intensity on T1 weighted-fluid-attenuated inversion recovery (FLAIR) in axial section although there was high signal intensity on T2 weighted images (Figure 1). The lesion had an area of hyperintensity focus on T1 -weighted images. It was considered as presence of intratumoral hemorrhage.

The lesion showed intense contrast enhancement axial and coronal on T1 weighted images by intravenous administration of a contrast media (Figure 2). MRI examination of whole spine with contrast showed no pathology. Tumor markers such as alpha-fetoprotein (AFP) and human chorionic gonadotrophin (βHCG) in the serum and cerebrospinal fluid were at normal range. Surgery was performed, because neurosurgical consultation concluded that the lesion should be completely removed. Histopathological examination of the solid mass revealed a germinoma. The patient received chemotherapy and then, she took radiation therapy regularly. After surgery, visual deficiencies of the patient due to removal of compression on visual pathways have improved. She received desmopressin for treatment of diabetes insipidus. Corticosteroids and levotiron were given to the patient for treatment of hypocortisolemia and hypothyroidism.

Discussion

Germinomas in suprasellar and pineal regions are considered to arise embryologically from a midline streaming of totipotential cells that occurs early in development of the rostral part of the neural tube.

However, development of the third ventricle may cause deviation of ectopic germ cells from midline, and the cells may appear in the basal ganglia and thalamus (7). Rarely, thalamus may involve the basal ganglia and thalamus bilaterally (8,9).

Germinomas are the most common germ cell tumors that also contain embryonal cell carcinomas, endodermal sinus tumors, choriocarcinomas, teratomas and mixed germ cell tumors (1). Local spread of intracranial germinoma within the brain and throughout the subarachnoid space cause synchronous lesions. Primary intrasellar germinomas are rare and usually developed as secondary intrasellar extension of primary suprasellar tumor may traverse the hypophyseal-hypothalamic axis. Douglas et al (10) reported a case of primary intracranial germinoma involving floor of the sella turcica, sphenoid bone and sinus. In the current study, there was no bone or sphenoid sinus invasion and spread to subarachnoid space. The lesion was located in the suprasellar region. Hence, it was named as a primary suprasellar germinoma. Because the optic chiasm and pituitary stalk were compressed by tumor, the patient had visual disturbance and diabetes insipidus significantly.

Although pineal germinomas have male predominance, suprasellar lesions affect both sexes equally (11). In the current study, the patient was female. Subarachnoid spread of primary intracranial germinoma is common. Therefore, before surgery, MRI of the whole spine is necessary for the determination of metastasis (9). In the present study, we observed that MRI examination of whole spine with contrast revealed pathology.

Douglas et al. (10) reported that they did not remove all of the solid sellar mass in their case. After surgery, the patient was only treated with radiotherapy of the sella and the sphenoid region chemotherapy was not administered to the patient in the study of Douglas et al. Recently, the combined approach of adjuvant chemotherapy and radiotherapy have been increasingly used, because the combined approach reduces the dose and/or field of radiotherapy and ultimately decreases the late effects of radiotherapy. Germinomas are exquisitely sensitive to radiotherapy and chemotherapy (12). In our case, total resection of the tumor was achieved successfully. After surgery, the patient was treated with the combined approach of adjuvant chemotherapy and radiotherapy. MRI findings of germinoma in the suprasellar region tended to be equal to that of gray matter for all pulse sequences (5). Moon et al. (13) reported that they observed slightly hypointense on T1-weighted images although they observed slightly hyperintense on T2-weighted images of intracranial germinomas. After i.v. administration of contrast medium, tumors showed strong and homogeneous enhancement in all patients. We observed in MRI findings of the current patient that the pattern and degree of contrast enhancement on T1- and T2-weighted images were similar to their images. In the current case, slightly hypointense on T1 weighted images and hiperintense on T2 weighted images were observed. MRI showed intratumoral hemorrhagic focus and it was hiperintense on T1 weighted images. Post contrast examination showed intense enhancement of axial and coronal on T1 weighted images. In addition, Moon et al. (13) stated that the tumors' diameter ranged between 1.2 and 4.5 cm in pineal or suprasellar lesions. In current study, the lesion was approximately 5 cm in diameter.

In conclusion, primary intracranial suprasellar germinoma should be considered the first pathology in the differential diagnosis of suprasellar lesions in children especially those presenting with signs of visual disturbance and diabetes insipidus. We wanted to emphasize the importance of an early diagnosis in order to reduce radiation damage, progression of the disease and careful follow-up of these patients.

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