



A Case of Pituicytoma Presenting with Blurred Vision and Hormone Deficiency

Görme Kaybı ve Hormon Eksikliğiyle Başvuran Pituisitoma Olgusu

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Abstract

Pituicytomas are very rare and benign primary tumors of the neurohypophysis or infundibulum. They originate from pituicytes and the clinical features vary depending on the location of the tumor. Immunohistochemistry has an important role in the diagnosis. There is no pathognomonic feature for pre-operative diagnosis. Diagnosis is generally established after surgery. We present this case report to emphasize the clinical features and good prognosis as well as unusual locations of pituicytomas that may be observed on magnetic resonance imaging. *Turk Jem 2015; 19: 115-118*

Key words: Pituicytoma, neurohypophysis, immunohistochemistry

Özet

Pituisitoma, hipofiz bezinin infundibulum yada nörohipofiz bölgelerinden kaynaklanan nadir iyi huylu bir kitledir. Pituisitlerden orjin alır ve klinik özellikleri bulunduğu lokasyona göre değişir. İmmünohistokimyasal boyamalar en önemli tanı yöntemidir. Operasyon öncesi tanı koyduracak patognomik özelliği olmaması nedeniyle, tanı genellikle operasyon sonrası immünohistokimyasal boyamalarla konulmaktadır. Biz bu olguyu nadir bir tanı olan pituisitomanın klinik özelliklerini, iyi bir prognosa sahip olduğunu ve karşımıza manyetik rezonans görüntüleme de atipik lokalizasyonlarda çıkabileceğini vurgulamak amacıyla sunduk. *Turk Jem 2015; 19: 115-118*

Anahtar kelimeler: Pituisitoma, nörohipofiz, immünohistokimya

Introduction

Pituicytomas are very rare and benign primary tumors of the neurohypophysis or infundibulum. They originate from pituicytes which are the specialized glial cells settled in the perivascular zones of the neurohypophysis (1). The clinical features of pituicytoma are similar to that of other pituitary tumors and depend on the location of the tumor. Furthermore, imaging exams can suggest pituitary adenoma. Pituicytoma may lead to pituitary hormone deficiency when it is located in the sella turcica, whereas suprasellar lesions usually present with headache and visual symptoms (2,3). Immunohistochemistry also has an important role in the diagnosis. The staining characteristics of the pituicytomas include strong immunoreactivity for S-100 protein and glial fibrillary acidic protein (GFAP), and lack of reactivity for epithelial membrane antigen (EMA). Neuroradiological findings are not pathognomonic, therefore, the diagnosis is generally established after surgery (4). Indirect evidence of posterior pituitary involvement on imaging is helpful in considering a diagnosis of

a pituicytoma (5). No recurrence occurs after total resection of pituicytoma (6,7). Herein, we report this case report to emphasize the clinical features and good prognosis as well as unusual locations of pituicytomas that may be observed on magnetic resonance imaging (MRI).

Case Report

A 33-year-old man presented with a 6-month history of blurred vision and headache. Physical examination revealed body weight of 72 kg, height of 175 cm, body mass index (BMI) of 23.5 kg/m², and blood pressure of 130/80 mmHg. Other system examination findings were normal. Laboratory findings were as follows: fasting blood glucose level: 85 mg/dl, triglyceride: 187 mg/dl (35-150), LDL-C: 124 mg/dl (0-135), HDL-C: 37 mg/dl (40-60), BUN: 12 mg/dl (7-20), creatine: 1.0 mg/dl. His hormone profile showed decreased growth hormone (GH), IGF-1 levels (IGF-1: 96 ng/ml, normal range: 140-405 ng/ml (at age 30-40 years) and mildly elevated PRL levels (42 ng/ml, normal: 3.4-29). GH level was <0.01 µIU/ml and we

confirmed the diagnosis of GH deficiency with insulin tolerance test (ITT) and glucagon stimulation test (GST). The peak GH values were inadequate, i.e. 0.87 μ IU/ml and 0.44 μ IU/ml after ITT and GST, respectively. Other pituitary hormones levels were: fT3: 3.5 pg/ml (2.3-4.2), fT4: 1.2 ng/dl (0.88-1.72), TSH: 0.8 μ IU/mL (0.57-5.6), ACTH: 15 pg/ml (0-46), cortisol: 13 μ g/dl (5-25), FSH: 6 mIU/ml (1.9-18.9), LH: 4 mIU/ml (1.7-9.6), and total testosterone: 457 ng/dl (134-625). 1-mg dexamethasone suppression test was performed and after the test, cortisol levels were suppressed (cortisol: 0.4 μ g/dl). MRI revealed a 26x16x18 mm mass with suprasellar component compressing the optic chiasm and involving both cavernous sinuses. The lesion was isointense on T1-weighted MRI and hyperintense on T2-weighted MRI and was homogeneously enhanced with gadolinium (Figure 1, 2).

Neurohypophysis was not found on T1-weighted images in its usual location and it was pushed to the suprasellar region (Figure 3, 4). GH deficiency was considered to be due to the sellar mass. As a result, a preoperative diagnosis of non-secreting pituitary macroadenoma was established, and the patient underwent transsphenoidal surgery. Postoperatively, he recovered well. Visual fields improved markedly, and visual acuity was normal. The immunohistochemical staining results are summarized in (Figure 5, 6). The pituitary tumor showed a diffuse positive staining for S-100 protein antibody (Figure 6) and negative staining for synaptophysin, and chromogranin A. Pathological analysis revealed pituicytoma. According to MRI findings, we assumed that this pituicytoma originated from the neurohypophysis, because the neurohypophysis was in the suprasellar region. We followed up the patient and no tumor recurrence (Figure 7) and no hormone deficiency were observed after a follow-up period of 24 months.

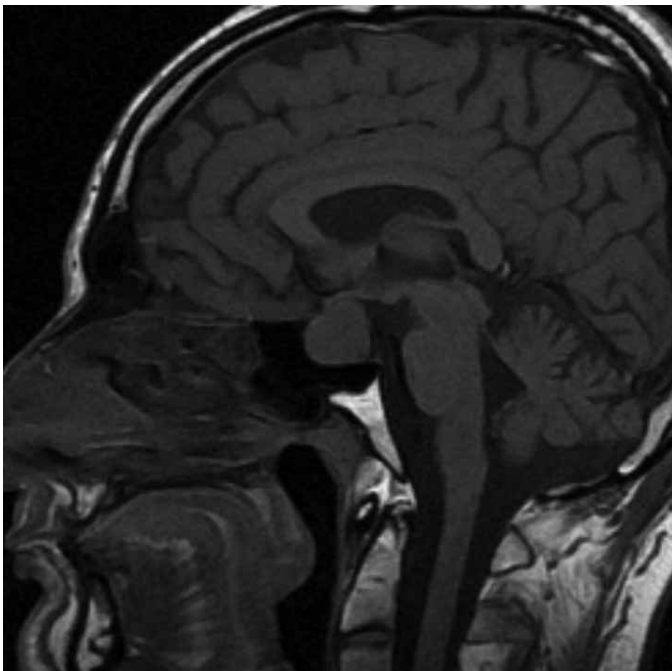


Figure 1. Sagittal unenhanced T1-weighted magnetic resonance imaging showing an isointense pituitary mass

Discussion

Liss and Kahn (8,9), first described the term 'pituicytoma' in 1958. In 2000, Brat et al. (10), defined specific pathological criteria for the diagnosis of pituicytoma, and in 2007, pituicytoma was included in the last World Health Organization (WHO) Classification of Central Nervous System Tumors to describe solid and circumscribed low-grade (WHO I) glial neoplasm that originates from the infundibulum or neurohypophysis, presumably arising from pituicytes (11).

The clinical presentation of pituicytomas is similar to that of other pituitary tumors, with patients typically presenting with visual or hormone deficiency symptoms. The sign and symptoms most frequently include headache, visual disturbances because of compression of the optic chiasm, and hormonal dysfunction due to mass effect on the pituitary gland and infundibulum (2,3). Our patient also presented with visual problems, increased PRL levels and decreased GH levels. High PRL levels were thought to be due to the stalk compression by the tumor. After surgery, the GH and PRL levels were normalized.

MRI showed a sellar and suprasellar lesion compressing the optic chiasm and involving both cavernous sinuses. The mass was solid, isointense on T1-weighted images and slightly hyperintense on T2-weighted images. The lesion presented intense and homogeneous contrast enhancement.

Neuroradiological findings are not pathognomonic, and the most common radiological preoperative diagnosis is pituitary adenoma. The radiological differential diagnosis should also include other sellar or suprasellar enhancing lesions, such as meningiomas, craniopharyngiomas, hemangiopericytomas, granular-cell tumors, pilocytic astrocytomas, gangliogliomas, sarcoidosis, germ-cell tumors, hamartomas, and metastatic

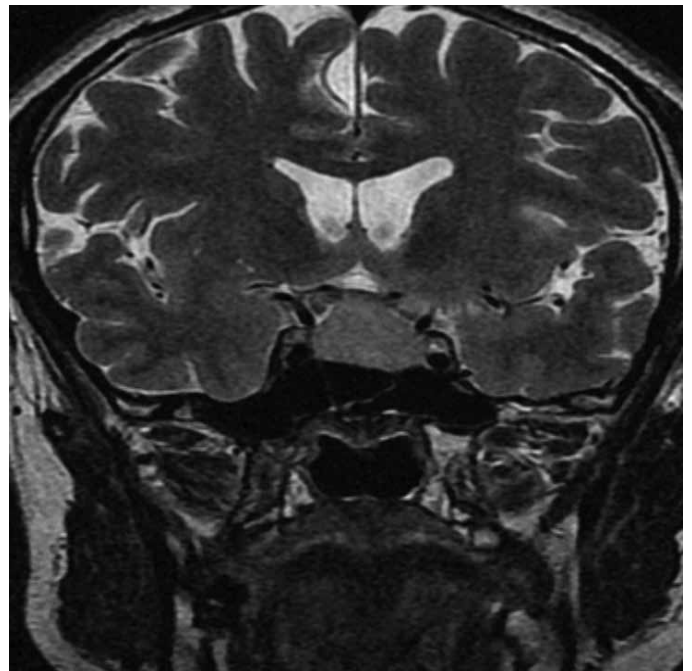


Figure 2. Coronal T2-weighted magnetic resonance imaging showing a slight hyperintensity of the tumor compared with the brain parenchyma

tumors (12,13). If the patient presents with acute hemorrhage, the differential diagnosis should also include Rathke’s cleft cyst, cavernous angioma, venous angioma, and aneurysm (14). Immunohistochemistry demonstrate S-100 and GFAP positivity, but negative staining for synaptophysin, chromogranin A and B in pituicytomas (15). The histological and immunochemical profiles distinguish pituicytomas from other lesions of the sellar and suprasellar areas. Pituitary adenomas are synaptophysin-reactive, frequently, chromogranin-positive and are negative for GFAP and

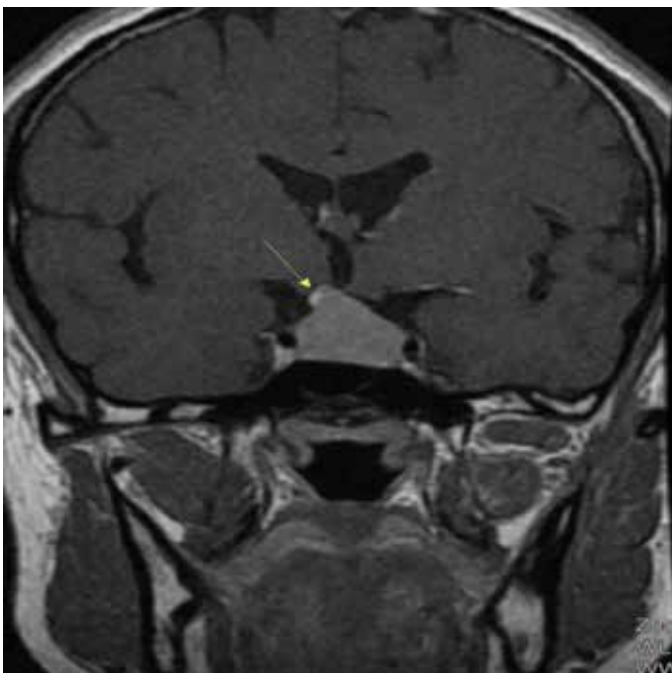
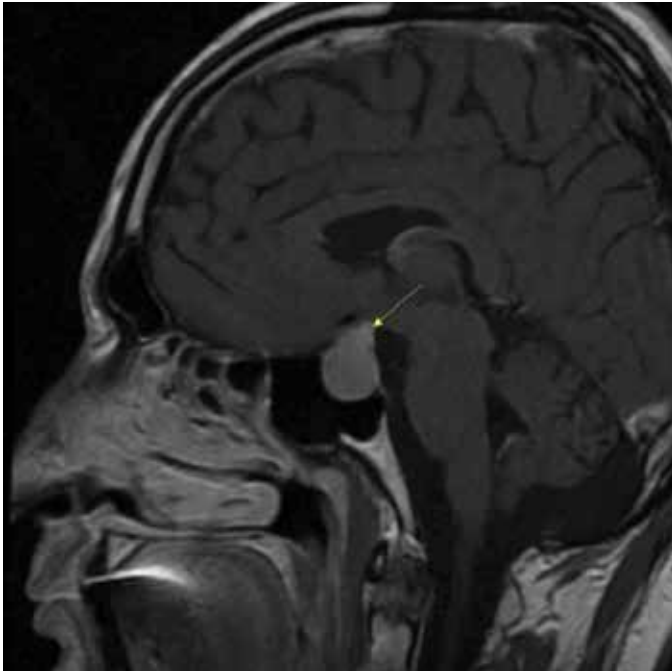


Figure 3-4. Sagittal and coronal contrast-enhanced T1-weighted magnetic resonance imaging demonstrating marked enhancement of the tumor. Neurohypophysis was pushed to the suprasellar region (arrow)

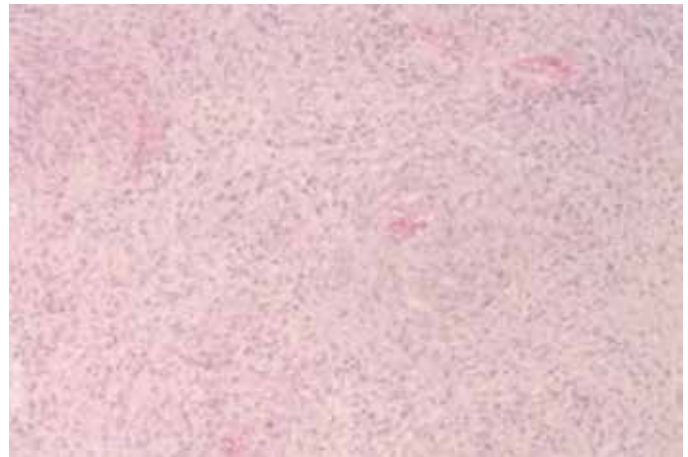


Figure 5. The tumor cells had oval-elongated nuclei and a large eosinophilic cytoplasm. There was perivascular pseudo rosette formation. Some of the tumor cells had pleomorphic appearance



Figure 6. Immunohistochemical stains showed that the tumor was diffusely positive for S-100

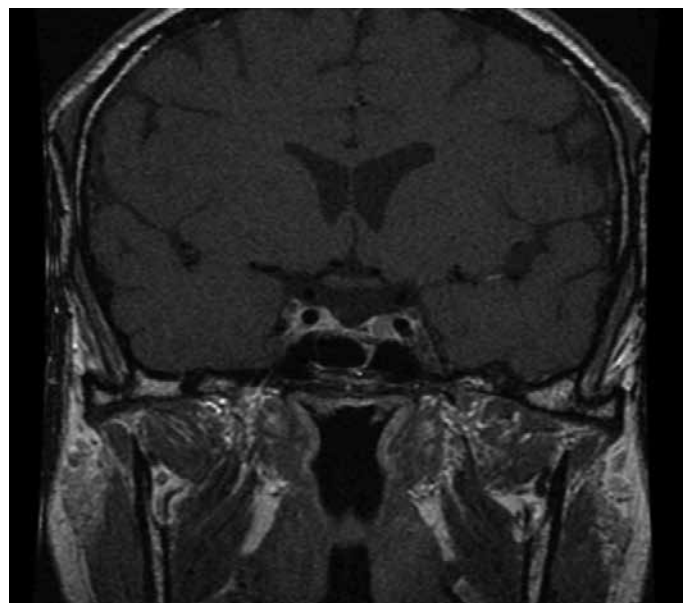


Figure 7. Post-operation images of the lesion on contrast-enhanced T1-weighted magnetic resonance imaging demonstrated no tumor

S-100. Immunohistochemical staining our case showed that the tumor was diffusely positive for S-100 and focally positive for GFAP. Furthermore, the resected tumor was immunonegative for EMA. Surgical resection is the mainstay of treatment for pituicytomas. In the largest series to date, Brat et al. (10) reported no recurrences of the tumor in 6 patients who underwent total surgical resection. We followed our patient for 24 months and there was no recurrence. In conclusion, we presented this case report due to its rarity. In addition, there was no recurrence of the pituicytoma and no hormone deficiency occurred during 24-month follow-up. We suggest evaluating patients in terms of pituicytoma in case of unusual location of an pituitary tumor in the neurohypophysis.

Informed Consent: Consent form was filled out by the patient.

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