Case Report
Pituicytoma, a rare sellar mass: a case report

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Abstract: We present the case of a 52-year old male with progressive reduction of vision. Though the case was initially misdiagnosed as pituitary adenoma by radiological studies and the intraoperative frozen section examination, the tissue sections were finally diagnosed as a rare sellar pituicytoma WHO grade I. The patient is recurrence-free for 2 years after the initial diagnosis. The histological features, immunoprofile and differential diagnosis are discussed in the current study.

Keywords: Pituicytoma, pituitary, immunohistochemistry, sellar

Introduction

Pituicytoma, arising from pituicytes of the neurohypophysis and infundibulum, is an extremely rare benign tumor of the sellar and suprasellar regions [1, 2]. It is previously known as “infundibulum” or “posterior pituitary astrocytoma”. The tumor usually presents as a solid, well-circumscribed, non-infiltrative mass [3]. The corresponding clinical symptoms are closely related to the location and the tumor size, including vision disturbance, headache, and hypopituitarism [4]. Clinically, the tumor mainly affects adult and only five cases occurred in children [5-8]. Neuroradiological findings are non-specific therefore the most common radiological preoperative diagnosis was pituitary adenoma. Histological diagnosis is still the most accurate examination for this tumor, especially with the help of immunohistochemistry (IHC) [1, 9, 10]. Till now, the best therapy is total resection. In the current study, we report a 52-year old male patient with a sellar mass and vision disturbance, with final histopathological diagnosis of pituicytoma.

Case presentation

A 52-year-old male patient was referred to our hospital with chief complaints of progressive decrease of vision for two months. A sellar mass was found by cranial computed tomography (CT) in the local hospital. No clinical features suggested endocrine abnormalities. Results from visual acuity tests showed acuity of 1.0 in the left and 0.5 in the right. No bitemporal haemianopsia were disclosed. Laboratory testing showed borderline decrease of plasma ACTH (6.75 pg/ml), serum FT4 (8.68 pmol/L), and severe reduction of serum testosterone (<0.69 nmol/L, normal: 6.27-26.28 nmol/L). These abnormalities may be caused by mass effect of the sellar tumor. Magnetic resonance imaging (MRI) revealed an enlarged pituitary gland and demonstrated a well-circumscribed sellar mass measuring about 2.3×3.1×2.1 cm in size. The lesion was isointense on T1- and T2-weighted images, and slightly enhanced following intravenous administration of Gd-DTPA. The optic chiasm was compressed by the lesion and slightly elevated. A nonfunctioning pituitary adenoma was suspected. A transsphenoidal surgery was carried out to remove the pituitary mass. During the operation, a small tumor was identified and removed. An intraoperative frozen section was examined and diagnosed as pituitary adenoma. However, one week after the surgery, the remaining tissue sections were given the final histopathological diagnosis of pituicytoma with evidences provided by IHC staining.
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Light microscopy showed multiple, small tumor fragments composed of plump spindle cells having eosinophilic cytoplasm, disposed in intersecting fascicles and bundles (Figure 1). There were no Rosenthal fibers, granular eosinophilic bodies, or Herring bodies. No nuclear atypia or mitotic activity was present. Tumor cells were diffusely labeled by vimentin, S-100, and TTF-1, with focal expression of glial fibrillary acidic protein (GFAP) (Figure 2A-D). The tumor cells were negative for epithelial membrane antigen (EMA), synaptophysin, CD68 and pituitary hormones (ACTH, HGH, and PRL) (Figure 2E-G). The index of Ki-67 was less than 1% (Figure 2H). Thus, other differential diagnoses such as granular cell tumor and pituitary adenomas were excluded with the assistant of IHC staining.

The patient is now in persistent complete remission 2 years after resection without evidence of or recurrence.

Discussion

Pituicytoma is defined as a benign spindle cell tumor originates from pituicytes, and was recognized as an entity by the WHO Classification of Tumours of the Central Nervous System in 2007 [2]. To data, there were no more than 80 cases of pituicytoma totally reported worldwide [5, 6, 9, 11, 12]. Only five cases occurred in children, all the others were in adult, and mainly in patients’ fifties and sixties, with equal distribution in males and females [13]. In most cases, pituicytoma mainly leads to vision disturbance, headache and pituitary insufficiency, such as reduction of libido, due to mass effect [9, 14, 15]. Only rare cases showed pituicytoma in association with hypercortisolism symptoms, and finally were proved the patient actually having asymptomatic pituicytoma and corticotropin-secreting pituitary adenoma simultaneously [5, 11].

Currently, there are no special patterns and characteristics can separate pituicytomas from the more common pituitary adenomas in sellar and suprasellar on MRI or CT scanning [3]. The radiological studies reveal iso- or hypointens T1 signals of lesions which are homogeneously contrast enhancing. These features are similar in patients with pituitary adenomas. Therefore, the radiology studies cannot provide accurate diagnosis in preoperative tests. To be differentiated from the more common pituitary adenoma or some other rare pituitary tumors, the tissues sections of pituicytoma can provide the most important evidence, especially when IHC are applied. Under the microscope, the tumor was composed of spindle-shaped cells arranged in sheet, storiform patterns or interlacing fascicles. The tumor tissue generally contains no Rosenthal fibers, eosinophilic granular bodies or Herring bodies [1, 16]. These assist in distinguishing the pituicytoma from pilocytic astrocytoma and normal neurohypophysis, especially when sampling is limited. The nuclei were oval or elongated and slight pleomorphic, with pinpoint nucleoli. Typically, atypia cannot be observed. According to the literature and our case, the tumor cells are diffusively positive for vimentin and S-100, focally positive for GFAP.
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Figure 2. The immunoprofile of the pituicytoma. Tumor cells were diffusively positive for vimentin (A), S-100 (B), and TTF-1 (C), with focal expression of GFAP (D), negative for EMA (E), ACTH (F) and CD68 (G). The index of Ki-67 was less than 1% (H), all at 200× magnification.

[1, 16]. Generally, the tumor cells are negative for EMA, synaptophysin, chromogranin A, and neurofilament. Since pituicytoma is not a hormone-secreting tumor, it is also negative for PRL, GH, FSH, TSH and ACTH. These can be helpful in differential diagnosis with pituitary adenomas. Tumor cells are also negative or variably positive for CD68, while granular cell tumor, another rare pituitary tumor, shows strong immunoreactivities to CD68 [16]. As a benign tumor, the index of Ki-67 in pituicytoma is low, mostly less than 2-3%. Several reports showed that pituicytoma is diffusively positive for TTF-1 in the cell nucleus, and this is consistent with our case [16, 17]. The reactivity to TTF-1 also helps to exclude pituitary adenoma in differential diagnosis.

In conclusion, our reported case demonstrates a pituitacytoma in a 52-year old male. The clinical examination, microscopic features and IHC results support the diagnosis of pituicytoma, a very rare pituitary tumor mainly occurring in sellar and suprasellar regions of adults.

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Disclosure of conflict of interest

None.

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