PITUICYTOMA; BENIGN SPINDLE CELL TUMOR OF POSTERIOR PITUITARY GLAND: A CASE REPORT

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ABSTRACT

Pituicytoma is one of the rare benign primary tumors of the posterior pituitary gland, originated from intrinsic special glial cells of the gland. The previous nomenclature of posterior pituitary tumors has been confusing with pituicytoma. "Pituicytoma" should be neurohypophyseal astrocytoma only. Clinical and laboratory presentations as well as neuroimaging are not specific; the correct diagnoses are achieved only after histopathological examinations. This article revealed a clinical presentation, neuroimaging findings and pathological features of a large sellar/suprasellar tumor in a 46-year-old male, having diagnosed of pituicytoma.

INTRODUCTION

Tumors occupying the sellar and suprasellar spaces are common, and their morphologic spectrum is well known. Most frequently encountered are pituitary adenoma, meningioma, craniopharyngiomas, germ cell tumors, gangliogliomas, harmatomas, epidermoids, lipomas and metastasis. The most common tumors found in the posterior pituitary are secondary metastases presumably due to the rich vascular supply.[1] Intrinsic tumors arising in the posterior pituitary gland are very rare. Pituicytoma is one of the rare, poorly characterized benign primary tumors of the posterior pituitary gland, originated from intrinsic special glial cells (modified astrocyte) of the gland. There were only four reported cases of this astrocytic tumors published during 1994 -2001. This case report contains clinical presentation, neuroimaging findings and pathological features of this type of tumor that may masquerade as the pituitary adenoma.

CASE REPORT

A 46-year-old Thai male was referred to the Prasat Neurological Institute, Bangkok for the evaluation of a sellar/suprasellar mass. He complained of 8-month history of progressive decreased vision that more pronounced on the left eye. He was recognized of narrowed visual field, especially on the temporal side. One-weak before admission, he complained of dizziness and blindness. He denied increased urination, diminished secondary sexual hair, weight gain or loss or galactorrhea. His medical history was unremarkable.

Physical examination on admission revealed a well-nourished man with no signs of hypophysial dysfunction. Neurological examination showed severe visual impairment of both eyes; finger count on the right and hand movement on the left. There were an afferent pupillary defect on the right and non-functioning on the left. Fundoscopic examination revealed left optic disc atrophy. The remainder of his neurological examination was normal.

Laboratory studies including electrolytes, blood glucose, urinalysis and a complete blood count were all within the normal limits. The endocrine studies

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for the pituitary gland were normal except for decreased serum level of morning cortisol to 0.58 mg/dl (normal 4.3 -22.4 mg/dl).

Preoperative computed tomography revealed a well-defined, midline mass in the sellar region with suprasellar extension. The tumor was solid, homogeneous attenuation and slightly denser than brain (fig 1). Homogeneously intense enhancement was noted after administration of contrast media (fig 2, 3). MRI of the brain with gadolinium revealed a large, sellar-suprasellar mass that displaced and compressed the optic chiasm and optic tracts, more on the left. The mass was hypointense on T1W image, had heterogeneously and moderately enhancement with gadolinium, and heterogeneous increased signal intensity on T2W image (fig 4, 5, 6). The tuber cinereum and stalk were not identified as well as non-visualized normal posterior pituitary bright spot. The radiological appearances could not distinguished from those of macroscopic pituitary adenoma.

The patient received 1-day preoperative intravenous hydrocortisone and underwent a right supraorbital craniotomy. The tumor was confined below right optic nerve and compressed and distorted right and left A1-ACA and left optic nerve. The olfactory nerve was also compressed by the tumor. Nearly total tumor removal was performed and the residual tumor was situated above the right optic nerve.

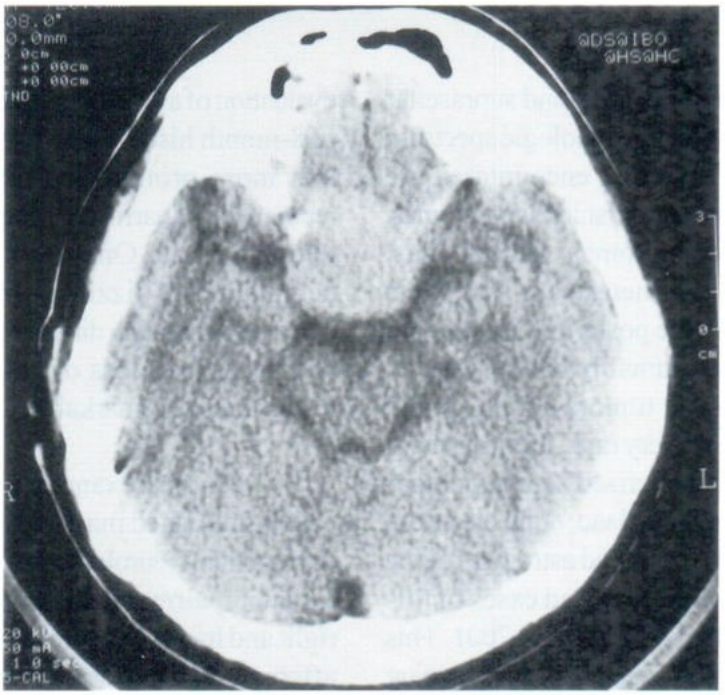
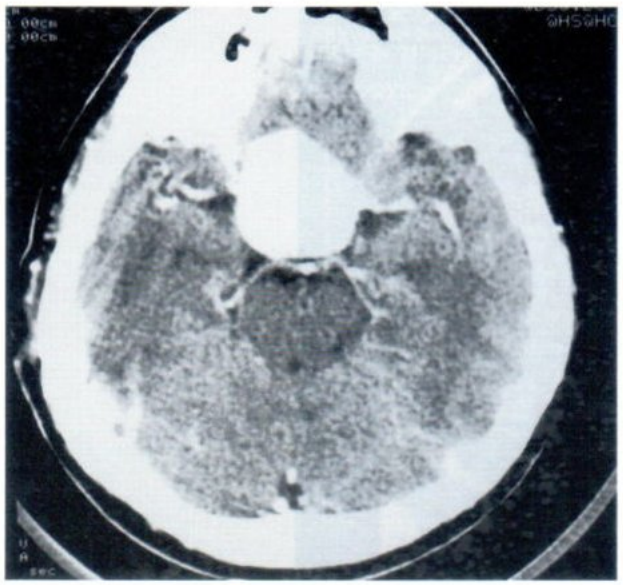


Fig 1 Non-enhanced axial CT scan shows midline homogeneous mild hyperattenuated-solid mass occupying within sellar region with suprasellar extension



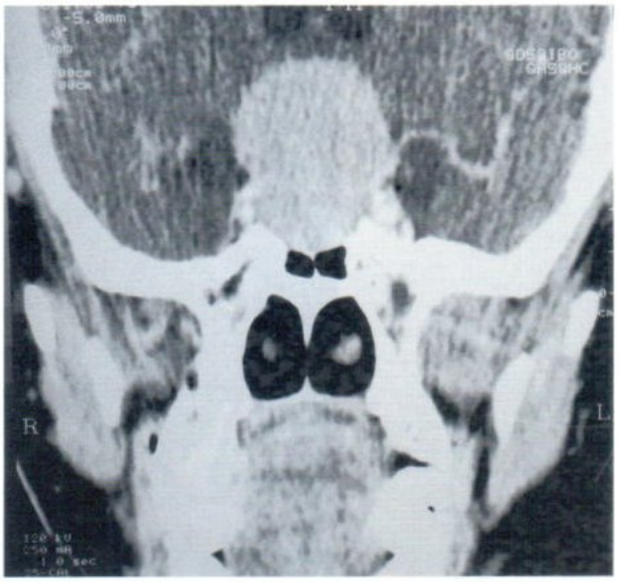
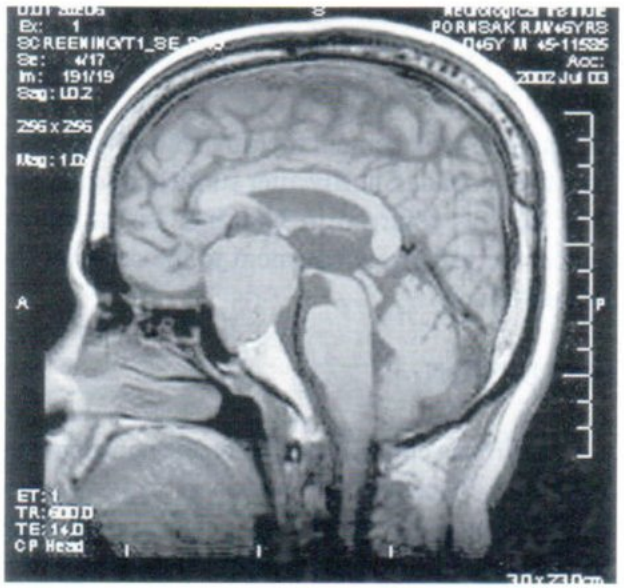


Fig 2 Axial and **Fig 3** coronal contrast enhanced CT reveals a large homogeneous intense enhancement to the mass that is occupying in a ballooning sellar turcica with suprasellar extension, giving a figure -of-eight in appearance.



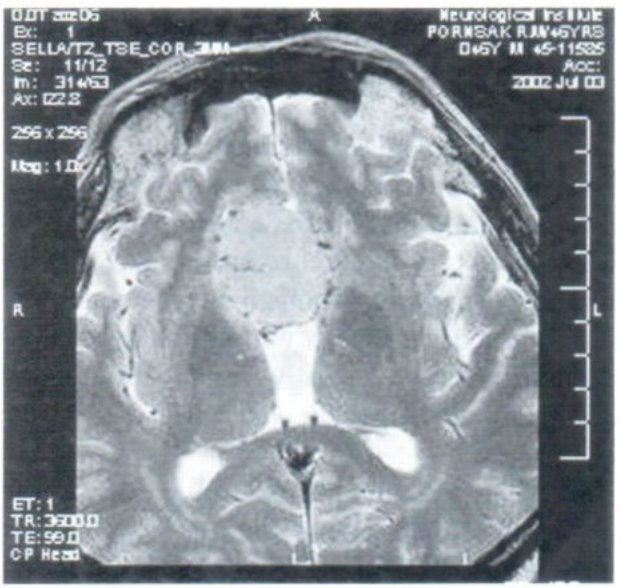


Fig 4 MRI sagittal SE T1 (TR/TE 600/14) in midline slice and **Fig 5** axial FSE T2W (TR/TE 3300/58) show a large sellar-suprasellar mass of relatively hypointense to gray matter on T1W and heterogeneous hyperintensity signal on T2W. The normal posterior pituitary bright spot is absent. The pituitary stalk could not be identified as the tumor size is large. Also noted on T2WI, there are prominent vascular signal void structures around the mass.

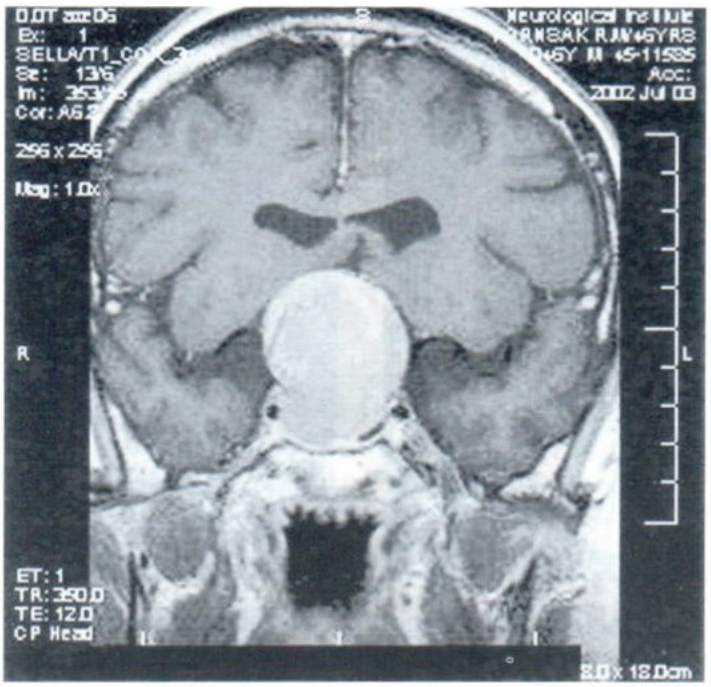


Fig 6 MRI coronal T1W (TR/TE 350/12) scan following intravenous gadolinium injection shows rather heterogeneously and modeately enhancement of the tumor mass.

On histological examination, the submitted tissue consisted of moderately cellular tumor. The tumor cells are medium in size, spindle shape and have round to ovoid, hyperchromatic nucleus with moderate cytoplasm. Mitotic figure, necrotic area, microcyst and granular body are absent. In addition, the tumor cells give a positive result with antibody against glial fibrillary acidic protein (GFAP).

The patient's postoperative course was uneventful. No episode of diabetes insipidus was documented. The hormonal assays in the fourth postoperative day showed slightly elevated serum level of morning cortisol to 2.4 mg/dl, but still lower than normal serum level (normal 4.3 -22.4 mg/dl). Decreased serum levels of LH2 and FSH were reported about 0.03 mIU/ml (normal 1.5 -9.3 mIU/ml) and 0.32 mIU/ml (normal 1.4 -18.1 mIU/ml), respectively. He was doing-well and receiving oral predisolone replacement therapy when discharged from the hospital. Shortly after surgical decompression, he was referred to National Institution of Cancer for radiation treatment to the sella for the residual tumor.

DISCUSSION

From a clinical standpoint, the nonadenomatous lesion in the pituitary region may give a radiological appearance identical to that of a pituitary adenoma and lead to misdiagnosis. Because intrinsic tumors arising in the posterior pituitary gland are very rare, the sellar region tumors are often thought to represent other more frequently occuring tumors such as pituitary adenoma, craniopharyngioma, germ-cell tumor or meningioma as well as other less common lesions such as metastatic carcinoma, schwannoma and chordoma.

The previous nomenclature has been confusing, the term "pituicytoma" have been used for a variety of tumors of the posterior pituitary[2] such as granular cell myoblastoma,[1] granular cell tumor, and choristoma.[3,4,5] The pituicytoma is most commonly found in older female people, with a sex ratio of 2:1 and in general appears after than the age of 30.[3,5] Few cases have clinical symptoms but most symptomatic tumors have been diagnosed as craniopharyngioma or meningioma before surgery.[4]

The clinical manifestations of the tumor of neurohypophysis or pituitary stalk are usually nonspecific such as headache and hypopituitarism. Compression of the optic pathways, causing loss of visual acuity, visual field defects (bilateral temporal field deficits) and optic atrophy, frequently occurs.[6,7] Mark suprasellar expansion may obstruct the third or lateral ventricles, causing signs of hydrocephalus or increased intracranial pressure.[8] Despite the involvement of the neurohypophysis and the pituitary stalk, diabetes inspidus is not a typical feature.[7] Clinical or laboratory signs of partial pituitary insufficiency or hyperprolactinemia, presumbly caused by the stalk effect.[8]

Anatomically, the neurohypophysis includes the posterior lobe of the pituitary (infundibulam process), the stalk or infundibular stem, and the median eminence of the tuber cinereum. The cellular elements that found in the posterior pituitary include pituicytes, microglia, portal blood vessels and terminal aborizations of the hypothalamic neurosecretory neurons. Pituicytes are considered to be modified glial cells.[1] On the current studies, most investigators consider pituicytoma to arise from pituicytes or Schwann cells of the posterior lobe of the hypophysis.[4] Hurler T, et all[1] as well as Rossi ML, et al[2,9] concluded that the term "pituicytoma" should be reserved for neurohypophyseal astrocytoma since the tumor are reviewed by some to originate from intrinsic glial cells of the gland, or the pituicytes.

There are only a few reports that clearly described the neuroimaging features of astrocytomas in the sellar region.[2,9] The tumor may occupy the stalk, posterior lobe or both. Computed tomogram show sharply-demarcate isodense or hypodense tumor with homogenous dense enhancement after administration of contrast medium. MRI also shows solid and partly cystic, discrete masses, which are usually isointense on T1W image, hyperintense on T2W images and which become hyperintense with contrast enhancement. If the tumor is large, the pituitary stalk and pituitary gland are not identified.[9]

Surgical removal by transphenoidal approach, which was associated with considerably less morbidity than was craniotomy, is the therapy for these tumors, as for other sellar lesions. The surgical goal should be limited to decompression of the suprasellar if total excision seems too hazardous.[10] Although curable by total excision, subtotal resection can be associated with recurrence.[11] Postoperative radiation therapy appears to further increase the mean survival time and extent time to tumor recurrence after subtotal resection.[8]

CONCLUSION

Such clinical appearances, laboratory datas and neuroimaging findings of the pituicytoma are not specific and may also be observed in other neoplasms of the sella, and probably because of their rarity, none have been diagnosed preoperatively.[5] Solid contrast-enhancing masses in the sellar region, are often thought to represent other frequency sellar region tumor such as pituitary adenoma. The correct diagnoses of these tumors are usually achieved only after histopathological examinations of tumor tissue. From the study of this case, the diagnosis of pituicytoma should be considered if the tumor is suspected to be originated in the posterior lobe of the pituitary gland.

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