

Lymphocytic infundibulo-neurohypophysitis mimicking a pituitary adenoma

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Abstract

A rare case of infundibulo-neurohypophysitis mimicking a pituitary adenoma is presented. A 69-years-old female patient developed polyuria and polydipsia. Laboratory analysis revealed central diabetes insipidus. No hormonal abnormalities. Cranial-magnetic resonance imaging (MRI) showed a left sided mass in the adenohypophysis presuming a pituitary adenoma. The mass had contact to both internal carotids. Admission to our department for neurosurgical treatment followed. Ophthalmologic examination and neurological examination yielded normal findings. A second MRI focussing on the sellar-region showed a left-sided (T2-MRI.hyperintense), distended adenohypophysis, without contrast enhancement in T1. The stalk appeared thickened. T1-weighted sequences of the neurohypophysis showed loss of signal intensity. We diagnosed an infundibulo-neurohypophysitis and abstained from surgical removal. The patient was discharged under treatment with corticosteroids and desmopressin. Hypophysitis is rare and shows special clinical characteristics. Despite defined radiological features to differentiate between hypophysitis and adenoma the possibility of misdiagnosis, and unnecessary surgical procedures, should always kept in mind.

Introduction

Lymphocytic hypophysitis (HYP) is a rare inflammatory condition classified in adenohypophysitis (aHYP), infundibulo-neurohypophysitis (nHYP) and panhypophysitis.¹ aHYP mainly occurs in women, often during pregnancy or postpartum period, causing hypopituitarism, while nHYP usually presents

with diabetes insipidus (DI). Diagnosis of HYP is based on endocrine dysregulation and abnormal findings on cranial magnetic resonance imaging (MRI).²

We present the case of a patient, who was admitted to our department with a suspected diagnosis of a pituitary adenoma, diagnosed as lymphocytic infundibulo-neurohypophysitis after more accurate neuroradiological and endocrinological investigations.

Case Report

A 69-years-old female patient suddenly developed polyuria and polydipsia. Laboratory analysis revealed a central complete DI. Her urinary osmolarity and plasma osmolarity were 70.5 mOsm/kg and 317 mOsm/kg respectively with oral free water intake. The peak plasma antidiuretic hormone (ADH) level measured by radioimmunoassay was 1.3 pg/mL when plasma osmolarity was 317 mOsm/kg. Serum sodium and potassium concentration were 144 mmol/L and 4.1 mmol/L. Urinary sodium and potassium levels were 108 mmol/L and 37 mmol/L respectively. Cranial MRI showed a left-sided adenohypophyseal mass featuring a pituitary adenoma (Figure 1A and B). The mass appeared to contact both internal carotid arteries without deviation of the stalk or connection to the optic chiasm. She was admitted to our department for neurosurgical tumour removal. On physical examination she showed no signs other than dry mouth and coated tongue. Ophthalmologic and neurological examination yielded normal findings. Her past history included an intracerebral hemorrhage in 1996 treated by evacuation of the hematoma, decompressive craniectomy followed by reconstructive cranioplasty. No evidence for recent head trauma or infections. Basal anterior pituitary function tests were normal except for low increase of Human Growth Hormone (18.10 ng/mL) which was probably stress-related. The clinical signs, the results of the laboratory data and the evaluation of MRI were not convincing upon the diagnosis of pituitary adenoma. We performed a second MRI focused on the sellar region. MRI showed a left-sided distended adenohypophysis with highly intense signal in the T2-weighting, without contrast enhancement in the T1-weighting. The stalk appeared thickened. Predominantly remarkable was the loss of signal intensity in the T1-weighted sequences of the neurohypophysis (Figure 1C). Combining our test results we drew the diagnosis of nHYP as our conclusion and abstained from surgical procedure. The patient was discharged and was treated with 50 mg corticosteroid twice a day and desmopressin spray. Six months later her condition was stable, but the symptoms were not declining.

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Discussion

aHYP is considered to be an autoimmune reaction in the anterior pituitary gland frequently associated with female gender, pregnancy or presenting during the postpartum period.³ However, according to different autopsy series occurrence of aHYP could also been shown in males.⁴ aHYP most commonly presents with symptoms of sellar mass with or without varying degrees of hypopituitarism, whereas DI is uncommon.⁵ nHYP is a distinct entity, first reported by Imura, in which inflammation is confined to the hypothalamic-neurohypophysial system frequently presenting with DI.^{2,6} DI is a clinical syndrome, characterized by polyuria and polydipsia, which can be divided in central (ADH deficit) and nephrogenic type (ADH-resistant). Central DI is usually caused by head trauma, infections, inflammations (e.g. meningitis, hypophysitis) or neoplasms (e.g. adenoma). Inflammatory processes of the hypophysis can be easily misdiagnosed because of their clinical and radiological features mimicking tumors in the sellar or parasellar region. MRI has improved the diagnostic accuracy by differentiating HYP from pituitary tumours.⁴

HYP usually presents as symmetrical enlargement of the pituitary gland while it is asymmetrical in patients with adenoma. The stalk is thickened but non-deviated while deviation of the stalk is observed commonly in adenomas. Patients with HYP show an intense and homogenous enhancement of the mass after gadolinium contrast and also a strip of enhanced tissue along the dura mater (dural tail) while patients with adenoma show a slightly delayed and inhomogenous enhance-

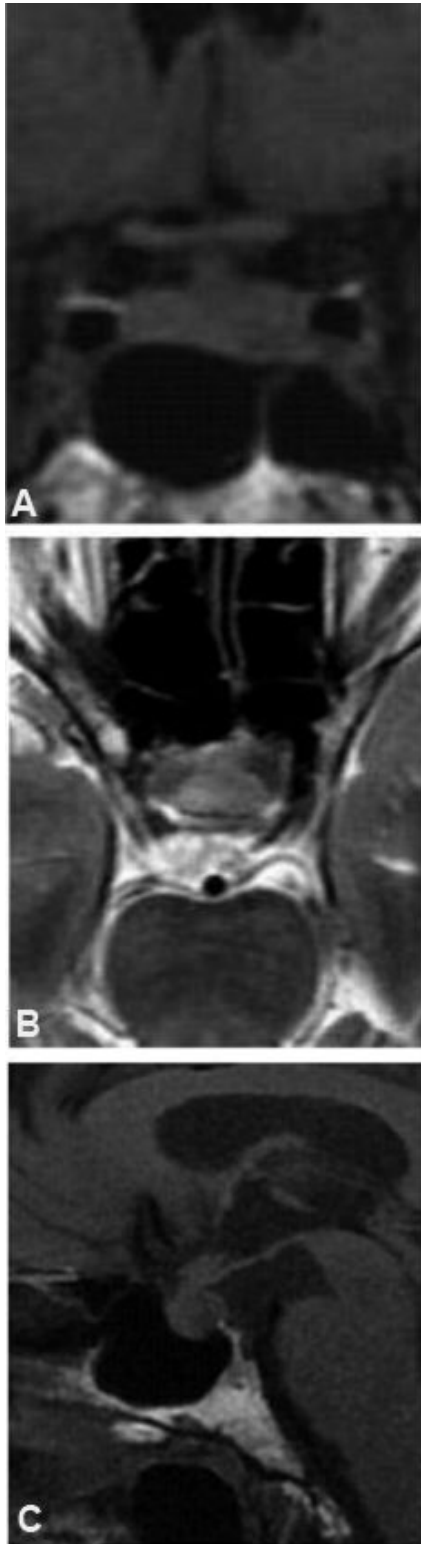


Figure 1. (A) T1 weighted magnetic resonance imaging: Coronal section of the pituitary showing a thick pituitary stalk. The hypophysis seems to be tumorous on the left side. (B) T2 weighted MRI: In the transversal section section an adenoma of the left pituitary gland is suspected. (C) On the sagittal section of this T1 MRI the neurohypophysis which is normally hyperintense is hypointense.

ment. The neuroimaging findings that could lead in particular to the diagnosis of nHYP are the absence of abnormalities in anterior pituitary gland, the thickening of the pituitary stalk and the enlargement of the neurohypophysis. Furthermore a signal loss of the posterior lobe on T1-weighted images is characteristic.² Apart from these schematic differences it is important to note that some variants of nHYP may form a mass not localized in the neurohypophysial space but involving the adenohypophysis, thus resembling adenomas and other tumours.^{7,8}

In these particular cases it is important to perform a high resolution MRI focussed on the sellar region to notice the thickness of the stalk and the signal intensity of the neurohypophysis, that are the two cardinal signs that can lead to the correct diagnosis.

In patients with the radiological findings mentioned above, presence of DI and normal anterior pituitary gland function, the diagnosis of nHYP should be considered.

If clinical and neuroimaging findings are typical of this disease, surgical intervention including biopsy should be avoided since it does not improve the diabetes insipidus and can also lead to further endocrine disturbance.⁹ Spontaneous remission of this disease can occur but a treatment with immunosuppressive drugs like glucocorticoids is generally recommended, even if this therapy seems to be effective in only about 30% of all treated patients.^{4,10,11} The patients should be examined in follow-up-examinations including MRIs closely to register the success of therapy and to avoid dysregulation of the endocrine system.

Conclusions

HYP is a rare constellation with special clinical characteristics. Despite defined radiological features to differentiate between HYP and adenoma the possibility of a misdiagnosis, and consequently unnecessary surgical procedures, is always to be kept in a neurosurgeon's mind.

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