CASE REPORT

Extensive spherical amyloid deposition presenting as a pituitary tumor

*Department of Internal Medicine, Division of Endocrinology and Diabetology, **Department of Pathology, ***Institute of Neuroradiology, ****Department of Neurosurgery, University Hospital of Zurich, Switzerland, *****Division of Endocrinology and Metabolism, Department of Internal Medicine, Taubman Center, University of Michigan, USA

ABSTRACT. A 71-yr-old man was admitted for further evaluation and trans-sphenoidal surgery of a pituitary tumor. He complained of impotence and decreased libido over a period of about 40 yr. Thirty-eight yr ago he was treated for bilateral gynecomastia with galactorrhea. Endocrinological investigation at presentation revealed only mild hyperprolactinemia and hypogonadotropic hypogonadism. Pituitary magnetic resonance imaging (MRI) showed a tumor up to 2.5 cm in diameter with infiltration of the sphenoid sinus and right cavernous sinus. The tumor exhibited a heterogeneous hyperintense signal on T1-weighted images and hypointense signal on T2-weighted images. Standard trans-sphenoidal surgery was performed and a brownish mass was found inside the sella, which was removed. Histological examination of the mass revealed extensive spherical amyloid deposits with strongly positive immunohistochemical staining for prolactin. Therefore, a prolactinoma with extensive spherical amyloid deposition was diagnosed. Extensive spherical amyloid deposition is a rare finding in prolactin-secreting pituitary adenomas. So far, characteristic radiological findings by MRI have been described only twice. Due to characteristic MRI findings, the diagnosis of extensive intrasellar amyloid deposition can be entertained pre-operatively. Trans-sphenoidal surgical resection is essential to confirm the diagnosis histologically and because of the potential lack of tumor shrinkage under dopaminagonist therapy in this type of prolactinoma.

INTRODUCTION

Extensive intrasellar spherical amyloid deposition is a rare finding in pituitary adenomas, with less than 20 cases reported up to today (1). Spherical amyloid deposition is almost exclusively encountered in PRL producing pituitary adenomas and only exceptionally occurs in GH and ACTH secreting or inactive adenomas (1). Abnormal processing of a hormone or prohormone by the adenoma cells has been suggested as the origin of the spherical amyloid formation (2). Amyloid deposits do not cause any characteristic clinical or biochemical features. Therefore, intrasellar amyloid deposition is not usually recognized pre-operatively. Magnetic resonance imaging (MRI) of intrasellar amyloid deposits have been described only twice (3, 4).

We report a patient who was admitted for further evaluation and trans-sphenoidal surgery of a pituitary tumor. Intra-operatively, parts of the sphenoid sinus and the sella were filled with a brownish waxy-like mass of soft consistency. The histological investigation of this mass revealed almost exclusively spherical amyloid bodies.

CASE REPORT

A 71-yr-old man was referred for further evaluation for an incidentally found pituitary tumor on a computed tomography (CT), which was performed due to a syncope. The patient had had impotence and decreased libido for about 40 yr, but attributed them
to psychosocial factors. Bilateral mastectomy due to gynecomastia and galactorrhea was performed 38 yr ago. Laboratory results from that time were not available. Arterial hypertension was treated with a β-blocking agent. Physical examination disclosed peripheral neuropathy with absence of ankle jerks and impaired vibratory sense. The rest of neurological examination was normal including normal visual fields. Testes were soft, 14 and 10 ml in volume. Skin was pale and showed fine wrinkles in the corners of the eyes and mouth. Body and facial hair was sparse, and pubic hair showed a female distribution pattern (Tanner P4-P5) suggesting hypogonadism. Laboratory investigations showed mildly elevated prolactin levels of 72.5 μg/l (normal 2.2-18.5). Total T was low at 2.5 nmol/l (reference 8.2-35) and free T was 7.2 pmol/l (reference 19-66 for men above 60 yr). FSH and LH serum levels were in the normal range (LH 2.4 IE/l, reference 2-12 and FSH 4.9 IE/l, reference 2-12). Thyroid indices were normal and serum cortisol level of 480 nmol/l (reference 280-690 nmol/l), drawn at 08:00 h confirmed a normal adrenal function. The IGF-I was low at 48 μg/l (reference for adults 100-300 μg/l). Routine hematological and chemical parameters including inflammatory markers were within the normal range.

Pituitary MRI demonstrated a sellar mass of 2.5 x 1.8 x 1.8 cm diameter, without suprasellar extension but with infiltration of the sphenoid sinus and right cavernous sinus (Fig. 1). The pituitary stalk was shifted to the left side. This tumor mass showed heterogeneous hyperintensity on T1-weighted images without iv contrast (Fig. 1A) and was hypointense on T2-weighted scans (Fig. 1D). After a trans-nasal/trans-sphenoidal access to the sphenoid sinus, the sphenoid mucosa was found to be normal, but the sellar floor was penetrated by a brownish, soft and wax-like, slightly dotted mass. Exploration of the sella revealed no classical pituitary adenoma. The tumor was re-

Fig.1 - MRI findings. Sagittal (A) and coronal (B) T1-weighted MR images without iv contrast showing the heterogeneously hyperintense mass (arrow). On coronal T1-weighted MR images with iv contrast, the mass is hypointense in comparison with the hypophyseal tissue (arrow) (C). On T2 coronal image (D) the mass is strongly hypointense as compared to the brain parenchyma.