Pituitary Adenoma Complicated by Hydrocephalus in a Patient Treated with Cabergoline

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Abstract

We describe a 55 year old, female patient with known pituitary adenoma since 9 years who was lost to follow up during 8 years. She presented with temporal vision loss, no other complaints. Imaging showed a large sellar mass which expanded suprasellar with severe anterior displacement of the chiasma opticum and extending into the third ventricle with significant obstruction of the foramen of Monroe, leading to a dilatation of the right lateral ventricle. Clinical significant tumour progression occurred under treatment with Cabergoline, leading to hydrocephalus. Patient underwent semi urgent endoscopic transssphenoidal resection of the tumour. Three months after surgery the patient was asymptomatic and imaging confirmed a complete resection. Hormone levels where in normal range on account of the substitution therapy. This case stresses the importance of follow-up in patients with pituitary adenomas.

Keywords: Pituitary; Adenoma; Hydrocephalus; Cabergoline; Endoscopic surgery

Background

Pituitary adenomas can be an incidental finding on imaging or can present with neurological manifestations as a consequence of the mass effect or as a syndrome of hormone hyper secretion and/or deficiency [1-3].

Visual field deficits/decreased visual acuity (63.9%) and headaches (50.9%) are common symptoms in these cases. In contrast with headaches, visual disturbances tend to correlate with tumour size [4].

Neurological symptoms are more common in non-functioning adenomas because these tumours do not secrete sufficient hormones to cause endocrine-type symptoms. As a consequence the diagnosis is delayed until the patient presents with headaches or visual changes. Hydrocephalus as a complication of pituitary adenoma is infrequent. In the literature there are only a handful of case reports concerning this complication [5-12].

Case

A 55 year old woman with known pituitary adenoma since 2005 was lost to follow up from March 2007 until February 2015 because of anxiety of the patient. The last control Magnetic Resonance Image (MRI) was performed in 2006, the scan showed a macro adenoma with dimensions of 2.8x2.5 cm with a cystic and haemorrhagic component. At the time, the patient had no complaints of headaches, visual disturbances or other symptoms. Cabergoline treatment was initiated in June 2005 with 0,5 mg/day which was gradually augmented till 1 mg/day (June 2006).

The patient received suppressive therapy with Cabergoline 1 mg a day during the 8 years of lost to follow up. She contacted the endocrinologist to plan a follow-up appointment by reason of complaints of temporal vision loss. The patient denied complaints of headaches, dizziness or gait disturbances. No complaints of frequent passage of large volumes of urine. A MRI of the pituitary and blood sample was planned.

Blood sample showed an elevated prolactin level of 188 µg/L, a depressed LH of < 0.2 U/L, a depressed FSH of 1.7 U/L and a cortisol deficiency (9.6 µg/dl).

The MRI showed a large sellar mass (3.1 cm × 2.3 cm × 2.6 cm) which expanded suprasellar with severe compression of the chiasma opticum and extending through the bottom of the third ventricle with significant obstruction of the foramen of Monroe, leading to a dilatation of the right lateral ventricle. The outflow obstruction of the right lateral ventricle resulted in transepimedylial migration of cerebrospinal fluid (CSF). The mass was hyper intense and partly iso-intense on T2, T1 showed a cystic and/or necrotic component.

Subsequently the patient was contacted to come to an urgent, combined consultation with the endocrinologist and neurosurgeon. A Compute Tomography (CT)-scan for neuro-navigation during surgery was executed. Three days after the MRI investigation, four hands endoscopic transssphenoidal pituitary adenomectomy was performed. After removal of the tumour we had an endoscopic view into the third ventricle with visualisation of the foraminae of Monroe with the plexus choroideus, Sylvia aquaduct and a view in the dilated right lateral ventricle (Figure 1).

Post-operative the patient was transferred to intensive care. Her stay in the Intensive Care Unit (ICU) was prolonged due to a bilateral pneumonia and liquor leakage. A lumbo-external drain was placed to control the liquor leakage. As the evolution was not favourable, a revision procedure with reconstruction of the sellar floor with cartilage and repositioning of the nasoseptal flap was performed with success. The patient receives substitution therapy with L-Thyroxine 100 µg a day, hydrocortisone (20 mg - 10 mg – 10 mg a day) and nasal Desmopressine spray.

Microscopic examination of the biopsies confirmed the diagnosis.

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of a non-secreting pituitary macroadenoma. One month post-operatively the patient was feeling good: no complaints of headaches, temporal vision improved with only discrete loss in the left eye. Blood analysis show good results of the substitution therapy which is continued (Hydrocortisone 10 mg - 10 mg - 10 mg a day) Table 1. Three months postoperatively a control MRI of the pituitary gland showed no arguments for residue or relapse tumour. The supratentorial ventricle system is widened and the chiasma opticum is descended into the sella turcica. Insuline tolerance test showed a cortisol deficiency, relatively high ACTH and a significant growth hormone deficiency. Consequently growth hormone substitution therapy was started with Somatropine 5,3 mg, 0,2 mg once a day.

Discussion

This patient was treated with a high dose of cabergoline (1 mg a day) for multiple years. Under therapy, tumour progression continued until presentation even though cabergoline treatment is associated with minor tumour shrinkage [13,14]. This case demonstrates the importance of follow up in patients with pituitary adenomas. The association between tumour growth and the development of complaints is weak. This tumour gained volume until it compressed the optic chiasm before the patient presented herself to the doctor. At the moment of presentation, the tumour had a cystic, necrotic component. Imaging showed a dilatation of the ventricular system, right lateral ventricle more pronounced than the left. At the right side we could see transependymal migration of CSF on MRI. She had no other complaints besides temporal vision loss, however with the images from MRI we would suspect more clinical symptoms. We assume that the patient came just in time and that every day of delay for surgery increased the risk of major complications following the hydrocephalus [15].

Lessons to be learned from this case are (I) the importance of regular follow up in patients with pituitary adenomas, (II) the weak association between radiologic findings and clinical symptoms in patients with pituitary adenomas and (III) that even large tumours can be removed through an endoscopic trans nasal transsphenoidal surgery.

References


