Hypopituitarism Caused by Carotid Cavernous Fistula

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Abstract

Here we report a 79-year-old woman who presented with a 7-day history of headache, nausea, vomiting, and was found to have proptosis and ptosis. Laboratory findings showed hyponatremia, hypocortisolism, secondary hypothyroidism and low follicle-stimulating hormone (FSH) and luteinizing hormone (LH) levels. CT angiography (CTA) showed a vascular lesion in sella causing a mass affect on the pituitary gland which proved to be a carotid cavernous fistula (CCF) by conventional angiography. The lesion was subsequently treated with coil placement and patient’s hyponatremia was successfully treated with corticosteroid and thyroid hormone replacement. Though rare, CCF should be considered in the differential diagnosis of sellar lesions. Also, in patients with CCF hyponatremia, hypotension or signs of hypothyroidism should warrant a work-up for pituitary function.

Key words: hypopituitarism, carotid cavernous fistula, CT angiography, hypothyroidism


Introduction

Carotid cavernous fistula (CCF) is a rare vascular malformation in the brain. It results from a direct connection between the cavernous part of the carotid artery and the cavernous sinus (1-4). CCF usually presents with proptosis, ptosis, ophthalmoplegia, chemosis, increased intraocular pressure and disc edema (5). CCF can lead to enlargement of the pituitary gland because of venous engorgement without causing any signs or symptoms of hypopituitarism (6). CCF can also cause pituitary hypoperfusion (7). Hypopituitarism secondary to giant carotid artery aneurysm (8, 9) has been reported but we report the first case of spontaneous CCF causing hypopituitarism.

Case Report

A 79-year-old woman presented with headache, nausea, vomiting and diplopia. Her symptoms started approximately one week before presentation. Her pupils were equal, round and reactive to light. Neuro-ophthalmologic evaluation showed that she had right-sided proptosis and ptosis due to pupil sparing right oculomotor nerve palsy and mild disk edema. She was hypotensive with systolic blood pressure in the range of 80-90 mmHg which was difficult to treat with intravenous fluid replacement.

Lab data showed that she was hyponatremic with sodium of 128 mmol/L. Morning cortisol level was 6.5 mcg/dL (normal range: 6.7-22.7 mcg/dL) and ACTH level was 4 picogram/mL (normal range: 9-52 pg/mL). FSH and LH were both abnormally low with the amount of less than 2 mIU/mL (normal more than 15 mIU/mL in post-menopausal woman). Prolactin level was within normal limit (12 ng/mL) as well as Insulin-like growth factor 1 (25 ng/mL). Her free thyroxine (fT4) level was 0.5 ng/dL (normal range 0.5-1.6 ng/dL) and TSH was 0.28 mcIU/mL (normal range 0.36-5.6 mcIU/mL). Her serum and urine osmolality were 253 and 322, respectively and her urine sodium was 135 meq/L. A complete blood count showed white cell count of 7.7 million/mL with 7% percent eosinophils. These lab data are summarized in Table 1.

CT/CT angiography of the head showed a left side cavernous carotid fistula (Fig. 1). Conventional angiography confirmed the diagnosis of left sided Borden type 1 direct dural cavernous carotid fistula. Fistula was treated with coil placement (Fig. 2). She was administered oral corticosteroids and levothyroxine sodium (Synthroid) with improve-
ment in hyponatremia (serum sodium eventually improved to 139 mmol/L).

**Discussion**

CCF can be either traumatic or spontaneous. Spontaneous CCF as observed in the current case is most common in young males and post menopausal females. CCF is also subdivided into four subcategories based on the type of connection between the carotid artery and cavernous sinus (1-4). Indirect fistulas account for 15% of intracranial vascular abnormalities and are divided into 3 subgroups based on the origin of their arterial blood supply being from dural, subdural or meningeal branches of the internal carotid artery. Direct fistulas are more common and usually are the result of a single connection between the carotid artery and cavernous sinus, without involvement of dural or subdural branches. The present patient had a direct fistula between the carotid artery and cavernous sinus which is the most symptomatic condition because of the highest blood flow from the carotid artery to the cavernous sinus. It was unusual because this type of fistula usually occurs in a post-traumatic condition or as a complication of surgery (1-4) while indirect types of CCF are usually seen in older women. The present patient developed typical ophthalmologic abnormalities expected in high flow CCF including proptosis, ptosis and conjunctival chemosis, however intraocular pressure was normal. In addition to the signs and symptoms related to CCF, this patient had evidence of panhypopituitarism based on the lab findings suggestive of secondary hypothyroidism (low TSH and low free T4), hypoadrenalism (low cortisol and low ACTH level, hypotension) and hypogonadism. This was likely due to the mass effect as seen in the CT/CTA of the head. We did not perform any stimulation test because of patient’s critical condition, but we think the diagnosis of hypopituitarism was highly likely because she had a low level of LH and FSH in postmenopausal age, and a low level of TSH and ACTH. Hyponatremia could be secondary to hypothyroidism and adrenal insufficiency; however it is difficult to distinguish between this cause and syndrome of inappropriate anti-diuretic hormone secretion (SIADH) or cerebral salt wasting (CSW). In this case, correction of hyponatremia after treatment with corticosteroids and thyroid hormones is suggestive of hy-
Mass lesions are one of the most common causes of hypopituitarism. Any mass within sella can cause pressure on pituitary cells and disturb their function. Gonadotropins are the hormones which are mostly affected by the pressure of a mass lesion. Pituitary adenomas, meningiomas (10), primary and metastatic malignant lesions (11, 12) and abscess (13) are among the most common etiologies of mass lesions in sella. Vascular lesions are less common but important in the differential diagnosis. Carotid artery aneurysm has been reported as a cause of hypopituitarism (8, 9). CCF can potentially be a source of pressure on the pituitary because of its location, however to the best of our knowledge this is the first report of spontaneous CCF causing hypopituitarism. Prior studies have shown either that CCF can cause enlargement of the pituitary gland without affecting its function (6) or that CCF can cause hypoperfusion of the pituitary gland (7). Spontaneous CCF is usually caused by a ruptured intracavernous aneurysm, fibromuscular dysplasia, collagen vascular diseases and atherosclerotic vascular disease. In the present case, considering the age of the patient, atherosclerotic disease is the most likely predisposing condition. It presents with proptosis, chemosis, diplopia, cranial nerve palsy (III, IV, VI) and central retinal vein occlusion (3-5). In this case, the fistula was direct which means that the high flow rate with high pressure goes directly from the carotid artery into the cavernous sinus. This high flow pressure can cause dilatation of the veins and a mass effect on the adjacent structures. Therefore, we speculate that in this case, direct mass effect was the cause of pituitary dysfunction. This speculation is further confirmed by radiologic evidence. Unfortunately, the patient was lost to follow-up and thus we were unable to follow her hormone levels.

We suggest that hypopituitarism be considered and investigated in patients presenting with CCF, especially if they have serum electrolyte abnormalities such as hypotremia. In addition, CCF should be considered in the differential diagnosis of a sellar mass.

The authors state that they have no Conflict of Interest (COI).

References
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