Interdisciplinary Neurosurgery: Advanced Techniques and Case Management 2 (2015) 120–122

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery: Advanced Techniques and Case Management

journal homepage: www.inat-journal.com

Case Reports & Case Series (CRP)

Carotid artery aneurysm resulting in myxedema coma ☆

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ARTICLE INFO

Article history:
Received 5 January 2015
Revised 5 March 2015
Accepted 5 April 2015

Keywords:
Internal carotid artery aneurysm
Myxedema coma
Pituitary apoplexy
Sellar mass

ABSTRACT

Intra-sellar aneurysms are a rare, but important consideration when evaluating pituitary masses. Identification of aneurysms is critical to appropriate treatment and avoiding perilous consequences. These vascular aneurysms can result in severe endocrine dysfunction due to mass effect, stripping of the vascular supply to the pituitary, or hemorrhage. Here we describe a novel case of spontaneous myxedema coma and pituitary apoplexy secondary to a large internal carotid artery aneurysm.

Introduction

Aneurysms with components extending into the sella turcica a rare, but important consideration when evaluating sellar and parasellar masses. Mistaking a calcified and/or partially thrombosed aneurysm for a tumor can have catastrophic intra-operative consequences. Severe endocrine dysfunction is uncommon, but may result from mass effect, vascular steal phenomenon, or hemorrhage. Here we describe a rare case of spontaneous myxedema coma and pituitary apoplexy secondary to a large internal carotid artery aneurysm.

Case report

A 78-year-old woman presented with symptoms of sudden polyuria, headache, and vomiting for 24 h. Bradycardia (52 bpm), hypothermia (33.9 °C) and hyponatremia (sodium 117 mmol/L) were present on initial assessment. She reported a history of a sellar mass initially diagnosed 24 years earlier. She had refused surgery and opted to monitor the lesion with serial imaging. Per the family and patient, her hormonal blood work was normal and the mass had been stable, not requiring treatment. She had a stable left homonymous hemianopsia for many years. A large mixed density sellar mass was confirmed on initial CT imaging and she was referred for urgent neurosurgical evaluation. Prior to evaluation, she became disoriented and hypotensive requiring hemodynamic support and intubation for airway protection. Exam was notable for Glascow coma scale of 9/15 (E3, V1, M5), minimal reactive pupils bilaterally, and delayed tendon reflexes at the biceps; no significant cranial nerve, motor or sensory deficits were identified. Formal visual fields were not testable. Her skin and mucosa were normal with minimal peripheral edema. She was empirically treated for myxedema coma and adrenal crisis due to presumed pituitary apoplexy. Intravenous levothyroxine (T4), liothyronine (T3), and hydrocortisone were initiated. Central hypothyroidism and central adrenal insufficiency were later confirmed with a low free thyroxine (0.4 ng/dL), low total T3 (76 ng/dL), low cortisol (0.7 μg/dL), inappropriately normal TSH (3.57 mIU/L) and normal ACTH (16.5 pg/mL), respectively. Prolactin was mildly elevated (56.2 ng/mL).

MRI (Fig. 1) revealed a 5 × 3.2 cm heterogeneous mass centered in the sella turcica extending into the left cavernous sinus with lack of flow void in the petrous and distal cavernous portions of the left internal carotid artery. The optic chiasm was poorly visualized due to significant displacement by the mass. The differential diagnosis included hemorrhage within a pituitary macroadenoma, other tumors including osteosarcoma, osteochondtoma, chordoma and metastasis, or giant aneurysm. Prior to neurosurgical intervention, CT angiography (Fig. 2) suggested a non-enhancing large thrombosed internal carotid artery aneurysm with rim calcification and surgical biopsy was cancelled. The patient's family declined further imaging characterization or surgical intervention.

Initial glucocorticoid replacement did not lead to clinical improvement. Only after her thyroid hormone pool was repleted to a normal free thyroxine level (1 ng/dL) did she improve clinically and wean from the ventilator to extubation. Despite her initial complicated hospital course, she was discharged home on oral thyroid and adrenal replacement therapy.

Discussion

Supraclinoid carotid aneurysms mimicking pituitary adenomas have been described previously [1]. These aneurysms comprise less than 10% of

☆ Disclosure Statement: The authors have nothing to disclose.

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http://dx.doi.org/10.1016/j.inat.2015.04.001

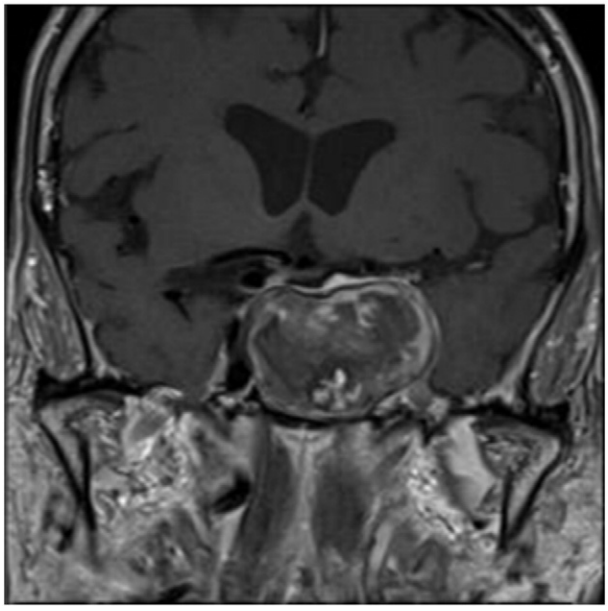

Fig. 1. T1 post-gadolinium MRI coronal image of large heterogenous sellar mass extending into the left cavernous sinus and compressing medial aspect of the left temporal lobe.

all cerebral aneurysms [2]. Para- and intra-sellar vascular lesions have been described across a broad range, including mycotic aneurysms, arteriovenous malformations and fistulas, cavernous malformations, and atherosclerotic lesions, mostly in case reports [1]. Advanced imaging modalities such as CTA, MRA and/or digital subtraction angiography (DSA) may be employed to reach the most likely diagnosis. Multiple cases have been reported where a parasellar or sellar aneurysm was discovered only at surgical exploration [3,4]. Lesion characteristics can be non-specific on CT scan. Non-contrast and post-contrast T1-weighted (T1WI) and T2 weighted (T2WI) MR sequences can be critical to evaluation of luminal flow and signal voids. These flow characteristics can differentiate patent lumens, partial or complete thrombosis. In a retrospective cohort of 17

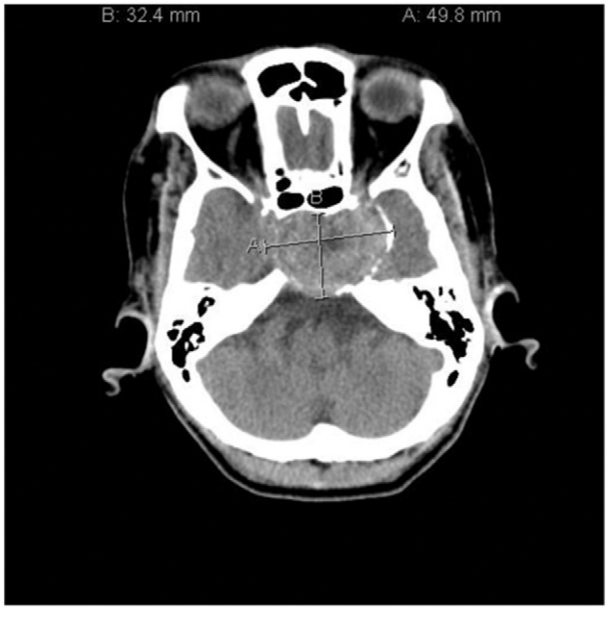

Fig. 2. Non-contrast CT sagittal image of peripheral calcification of high density sellar mass. Carotid aneurysm was highly suspected when combined with markedly diminished flow on CTA.

cases of giant intracranial aneurysms, a flow void sign on T2WI (sensitivity of 88%, specificity 100%), enhancement on contrast-enhanced T1WI (sensitivity 89%, specificity 100%), and presence of either flow void or enhancement on contrast-enhanced T1WI (sensitivity 100%, sensitivity 100%) indicated luminal patency. Eighty percent of those aneurysms characterized had an "onion skin" appearance on T1WI [5]. Ultimately, conventional angiography (CTA, MRA) is required for diagnosis and management [2].

Management of aneurysms proximal to the carotid bifurcation has evolved significantly over the past twenty years. Initially, aneurysm clipping, wrapping, and vessel bypass dominated the surgical treatments. More recently, endovascular options and technologies have emerged as complementary and less invasive, effective techniques. These include coiling, stenting, and flow diversion-based approaches. The decision to treat depends on numerous factors related to patient age and co-morbidities, aneurysm size, morphology, and location, and rupture status. In this case, there was no evidence of aneurysm rupture and the patient improved clinically with hormone replacements. As noted above, she and her family declined additional imaging and interventions.

Large aneurysms are infrequently associated with pituitary deficiencies and have been mistakenly diagnosed as prolactinomas in some reported cases [3,6]. In a retrospective review of over 4000 patients with a diagnosis of hypopituitarism, seven cases were secondary to an intrasellar aneurysm (0.17% prevalence). All 7 cases were associated with adrenal insufficiency, followed by 6/7 with thyroid and 5/7 with gonadal dysfunction. In the setting of a large sellar mass, decreased hypophyseal portal vein blood supply likely leads to reduced tissue perfusion, areas of ischemic necrosis and potentially decreased hypothalamic hormone stimulation to the anterior pituitary [7]. Select case reports have described short stature and diabetes insipidus [1,6]. Hyperprolactinemia is most likely from mass effect and posterior pituitary stalk compression rather than lactotroph hypertrophy.

Acute pituitary tumor apoplexy in the setting of a pituitary mass is a rare event, albeit even more rare in the setting of a sellar aneurysm [4]. Sudden bleeding increases intrasellar volume and pressure resulting in sudden headache, visual changes and often pituitary dysfunction. In this case, pituitary apoplexy was likely secondary to pituitary infarction via compression of the vascular blood supply rather than spontaneous hemorrhage into the pituitary itself. ACTH deficiency is the most common and most clinically relevant hormone deficiency [7], however, in this patient's case, identification and treatment of central hypothyroidism were critical factors to improving her clinical condition.

Prior to hospitalization, this patient had no symptoms of pre-apoplexy hypothyroidism and no known biochemical evidence of thyroid hormone deficiency. She denied a history of weight gain (54 kg, BMI 20 kg/m^2), constipation, fatigue, exercise intolerance, cold intolerance, coarse or dry skin or muscle cramps. She underwent normal age-related menopause. Her initial diagnosis of a sellar mass and subsequent care was completed out-of-state and confirmatory labs are not available, however, the patient and family maintain that she was in her usual state of good health and had no hormone abnormalities prior to presentation. The significant hyponatremia, we suspect, was secondary to cortisol and thyroid hormone deficiency, in which there are impaired ability to excrete a dilute urine and impaired delivery of salt and water to the diluting sites of the nephron [8].

Therefore, it is presumed that the acute exam findings of hypothermia, hypoventilation, bradycardia, hypotension, depressed consciousness and laboratory findings of hyponatremia and low free thyroxine and low total T3 were consistent with a clinical diagnosis of myxedema coma. Her polyuria may be explained by her hypokalemia on presentation (K 2.6) which has been associated with decreased collecting tubule responsiveness to antidiuretic hormone (ADH). Her polyuria and hypokalemia resolved spontaneously with no clinical or biochemical evidence of diabetes insipidus.

Use of thyroid function testing alone can be difficult when the etiology is of a central pathology as labs can look similar to those in non-thyroidal illness (i.e., a variable TSH and free T4, and low total T3 — all dependent on the duration of disease). In this case, it may have been useful to obtain a free thyroxine by equilibrium dialysis, using high-pressure liquid chromatography and tandem mass spectrometry, which is unaffected by changes in binding proteins as in the setting of critical illness.

In our patient, the myxedema coma diagnostic score was 110 with >65 being highly suggestive of myxedema coma [9]. Myxedema coma typically occurs after an inciting event such as infection, trauma, MI, CVA, CHF, or medication non-adherence which was not present in our patient. It is most commonly associated with primary hypothyroidism and rarely associated with central or secondary hypothyroidism (4/23 and 0/21 in two case series) [9,10]. The association of myxedema coma with a cerebral artery aneurysm has not been previously described to our knowledge.

Although there is variation in the treatment of myxedema coma, we preferred to aggressively replete her peripheral stores with a combination of T3 and T4. Treatment with T3 offers a more rapid onset of action compared to T4 and is the more bioactive thyroid hormone. In addition, conversion of T4 to T3 can be impaired in severe hypothyroidism. T3 therapy was discontinued after 3 days and T4 oral therapy was continued at weight based dosing.

Conclusion

Vascular aneurysms are rare sellar or parasellar masses, but their identification is critical to appropriate treatment. Evaluating the intraluminal blood flow on MRI prompted subsequent angiography and should be considered when there is suspicion that a vascular lesion may be mimicking a sellar mass. Prompt complete pituitary hormonal evaluation should be undertaken, as pituitary deficiencies have been described. Rarely, potentially devastating hormonal deficiencies could be present or evolve in the setting of pituitary apoplexy from a carotid artery aneurysm including myxedema coma.

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