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HYPOPITUITARISM AND AMENORRHEA-GALACTORRHEA SYNDROME CAUSED BY THROMBOSIS OF BOTH INTERNAL CAROTID ARTERY AND GIANT INTRASELLAR ANEURYSM

Case report

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ABSTRACT - Giant intra and parasellar aneurysm with a spontaneous thrombosis of internal carotid artery is rare. We report the case of a 34 years old woman presenting a unique giant sellar and parasellar aneurysm associated with hypopituitarism and amenorrhea-galactorrhea syndrome. Computed tomographic scans and magnetic resonance images were suggestive of a sellar tumor with a cystic component. Digital cerebral angiography showed spontaneous thrombosis of a intrasellar and parasellar carotid artery aneurysm and left internal carotid artery in the neck. A transseptal endoscopic biopsy was done and confirmed a thrombosed aneurysm. No other surgical treatment was required in this patient but permanent endocrinological treatment was necessary.

KEY WORDS: intracranial aneurysm, pituitary, prolactin, suprasellar mass, thrombosed aneurysm, endoscopy.

Hipopituitarismo e síndrome amenorreia-galactorreia causados por trombose de aneurisma carotídeo e da artéria carótida no pescoço: relato de caso

RESUMO - Aneurismas selares e paraselares gigantes com trombose da artéria carótida cervical e intracraniana são raros. Apresentamos o caso de mulher de 34 anos apresentando hipopituitarismo e síndrome amenorréia-galactorréia. A tomografia computadorizada craniana e a ressonância magnética foram sugestivas de tumor selar com componente cístico. Angiografia digital cerebral mostrou aneurisma único gigante selar e parasselar e trombose completa da artéria carótida comum esquerda no pescoço. A biopsia transeptal endoscópica da lesão mostrou tratar-se de aneurisma carotídeo trombosado. Não foi realizado nenhum outro tratamento cirúrgico na paciente, mas apenas tratamento endocrinológico de reposição.

PALAVRAS-CHAVE: aneurisma intracraniano, hipófise, prolactina, aneurisma trombosado, endoscopia.

Giant carotid aneurysm projecting into the sellar and suprasellar region may mimick a pituitary tumor and be responsible for mass effect symptoms dominating the clinical presentation[1]. A mass lesion in the sellar, parasellar and suprasellar areas can make a hypothalamic or pituitary stalk compression causing interference with the delivery of releasing and inhibiting factors to the pituitary, and destruction of pituitary tissue by an expanding mass lesion. Usually a combination of these mechanisms is present[2].

The prevalence of intracranial aneurysm ranges from 0.4% to 3.6% in autopsy studies and from 3.7% to 6.0% in studies of patients undergoing cerebral angiography[3]. Aneurysm projected into the sellar region account for 1% to 2% of all intracranial aneurysms[4,5]. To the best of our knowledge, this is the first case report of spontaneous thrombosis of both internal carotid artery and giant intrasellar aneurysm, with panhypopituitary and amenorrhea-galactorrhea syndrome.

CASE

A 34 years old woman was referred because of headache and amenorrhea-galactorrhea syndrome for 6 months. She was a non smoking woman and had no oth-

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er health problem. Examination revealed a conscious patient without neck stiffness. She had normal right visual acuity and no funduscopic abnormalities but decreased visual acuity on the left. The blood pressure was 130/80 mmHg. Pubic and axillary hair were absent, and there was a spontaneous galactorrhea. Basal endocrine studies showed a serum prolactin 75.50 ng/mL (normal range 1.9 - 25), TSH 0.212 mUI/mL (normal range 0.4 - 4.0) with T4 3.4 (normal range 4.5 - 12 mcg/dL), FSH 0.19 UI/mL (normal range 21.7 - 153), ACTH 8.8 pg/mL (normal range 12 - 70), LH 0.10 UI/mL (normal range 11.3 - 39), and GH 0.20 ng/mL (normal range 0.06 - 5.0). A computed tomographic (CT) scan (Fig 1) showed a large intra- and left parasellar homogeneous space occupying lesion with ring calcification. The lesion was not enhanced after intravenous contrast injection. Magnetic resonance image (MRI) was performed and T1-weighted images showed an isointense left paraselar lesion, surrounded by a high intensity ring, measuring 4.1 X 2.2 cm (Fig 2,3,4). After a gadolinium injection, there was no change of the lesion. A bilateral digital cerebral angiography showed a spontaneous thrombosis of internal left carotid artery in the neck, and there was no enhancement of the lesion by right carotid or vertebral injection, but only the indirect signs of supra and parasellar mass. (Fig 5) For diagnosis confirmation the patient was submitted to a minimally invasive biopsy by neuroendoscopic endonasal transseptal approach. Histopathology confirmed aneurysm wall with thrombus component. No other surgical treatment was envisaged and replacement endocrine therapy for life was initiated.

DISCUSSION

The present case reports the rare association of spontaneous thrombosis of a giant intra and left parasellar carotid artery aneurysm and the left internal carotid artery in the neck in a patient presenting panhypopituitary and amenorrhea-galactorrhea syndrome.

Carotid artery aneurysm in the sellar region mimicking pituitary tumor are uncommon. Extension into the sellar region with chiasmal compression was found as early as 1889 by Mitchell cited by Verbalis, et al.[2], and in 1912 by Cushing[6]. They can account up to 10% of the lesions causing parasellar syndrome[1]. Raymond[7] found 1.4% to 5% of all intracranial aneurysm referred to neurosurgeons projecting into the sella. On the other way there is a coexistence of aneurysms in 6.7% of pituitary adenomas[8,9].

The internal carotid artery thrombosis is asymptomatic in 27% of patients[3], and frequent spontaneous causes are atherosclerotic lesion, fibromuscular dysplasia and carotid arterial dissection. There is a high association between fibromuscular dys-

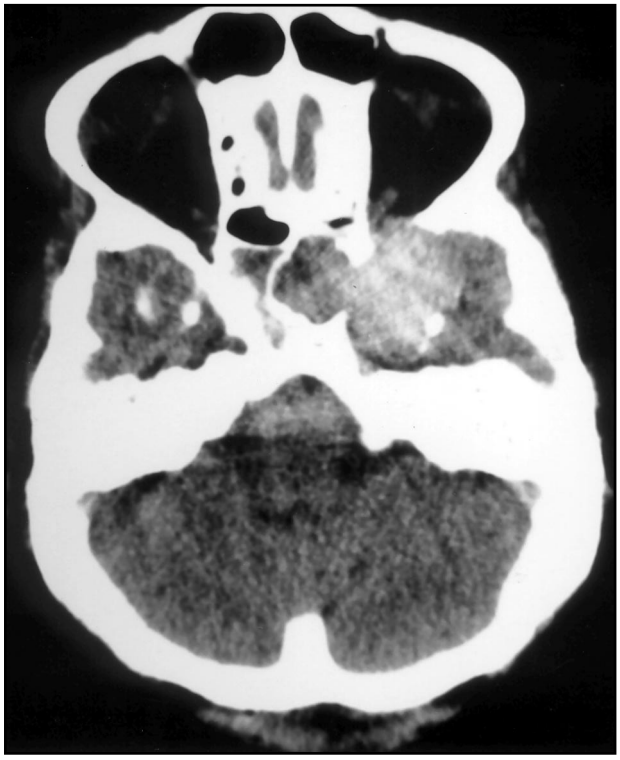

Fig 1. CT scan showing a sellar and parasellar homogeneous space occupying lesion with a ring calcification.

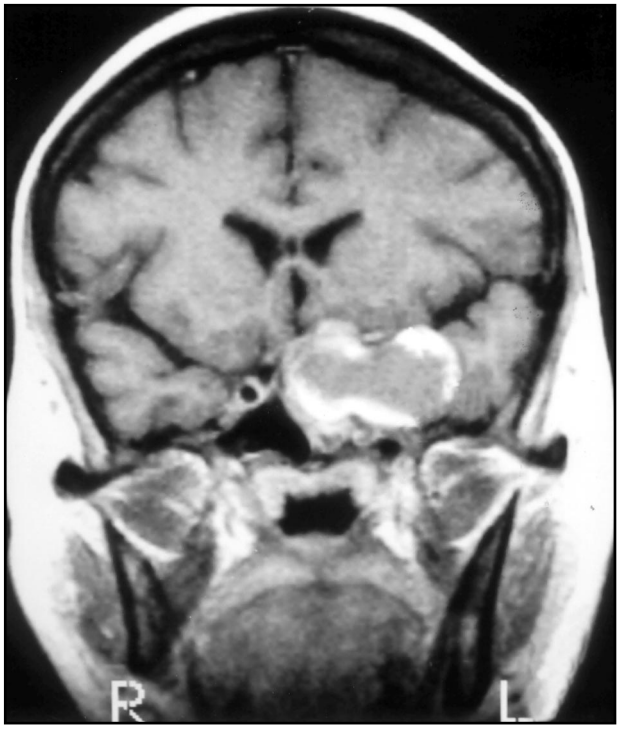

Fig 2. Coronal view T1-weighted MRI showing a iso and high intense T1-weighted mass in the sellar and left parasellar region, measuring 4.1 X 2,2 cm.

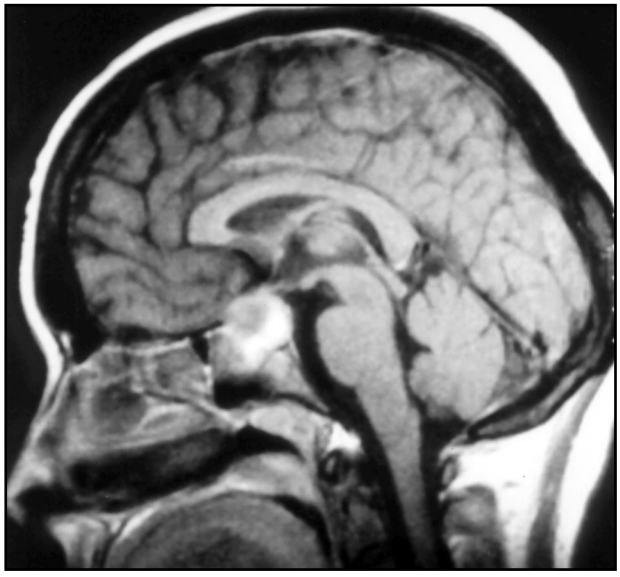

Fig 3. Sagittal view T1-weighted MRI showing a iso and high intense T1-weight mass in the a intra and supra sellar region.

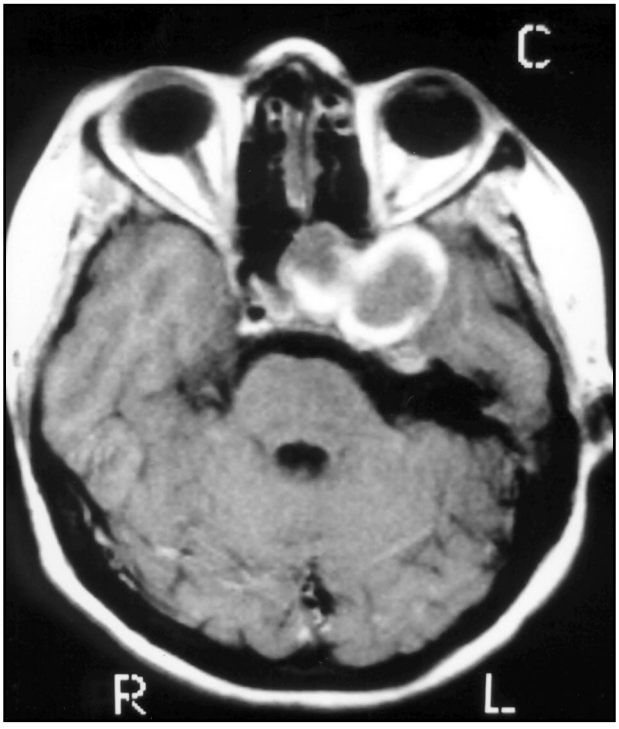

Fig 4. T1-weighted axial section demonstrating a iso and high intense T1-weighted mass in the sellar and left parasellar region.

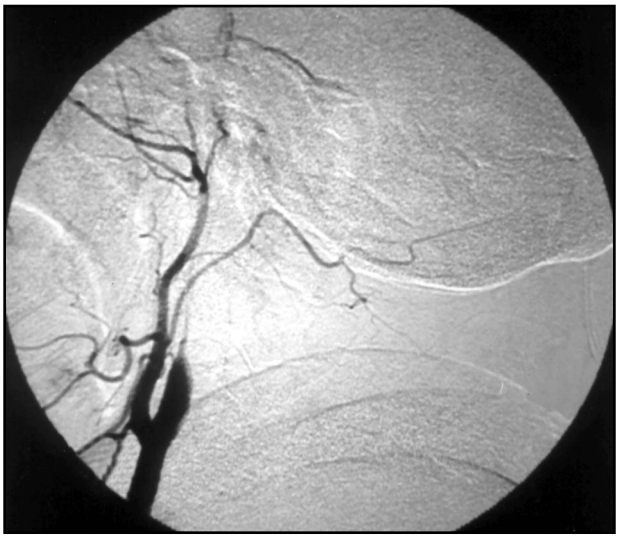

Fig 5. A digital cerebral angiography demonstrating a thrombosis of internal left carotid artery in the neck.

plasia and intracranial aneurysms estimated in 20 to 40%[10]. The histopathological exam of our patient was negative for this pathology. Although spontaneous thrombosis of a giant intracranial aneurysm is relatively common, occlusion of its parent artery is rare, the possible mechanism of carotid artery occlusion in this patient is due first to stretching of the internal carotid artery by the enlarged aneurysm, followed by compression of the internal carotid artery by the aneurysm itself. Next, the anterior clinoid process and the optic nerve were involved, and, finally, thrombosis of the aneurysmal cavity extended into the internal carotid artery itself[11].

The hypothalamus exerts a predominantly inhibitory influence on prolactin (PRL) secretion through one or more PRL inhibitory factors that reach the pituitary by way of the hipothalamic-pituitary portal vessels. PRL-releasing factors exist as well[12]. Disruption of the pituitary stalk leads to a moderate increase in PRL secretion and can show a decrease in secretion of the others pituitary hormones. The pathogenesis of endocrine dysfunction in patients with an aneurysm in the sellar and suprasellar area are: a) hypothalamic or pituitary stalk compression causing interference with the delivery of releasing and inhibiting factors to the pituitary, and b) destruction of pituitary tissue by an expanding mass lesion. Often a combination of these mechanisms is present, and the relative contribution of each to the pituitary dysfunction may change with continued growth of the mass[2].

Two pathogenesis possibilities can be found in hyperprolactinaemia. In the primary form the hormone is produced and secreted by a tumour mostly of the pituitary gland; in the secondary way, elevation of PRL levels might be caused by compression or distortion of the hypothalamus and stalk by a broad spectrum of different alterations like tumours (craniopharyngiomas, basilar meningio-

mas, ectopic pinealome, 3rd ventricle tumors, dysgerminomas, nonsecreting pituitary adenomas), infiltrating lesions such as sarcoidosis and eosinophilic granuloma, empty sella, lymphocytic hypophysitis, and aneurysm[12]. In secondary hyperprolactinaemia the PRL values usually do not exceed 130 ng/mL[3], in our patient it was 75.50ng/mL. Higher levels indicate PRL-producing adenomas[12]. Riedel et al.[3] demonstrates that a remarkable part (31.6%) of the sellar lesions can cause a secondary hyperpolactinaemia.

The most frequent etiology of panhypopituitarim is a pituitary adenoma. An exceptional cause, with less than 40 cases in the literature, is intrasellar aneurysm. The prevalence of hypopituitarism due to an aneurysm is 0.17%[5]. In the literature, pituitary dysfunction caused by intrasellar aneurysms shows that gonadal axis is deficient in 67.5% of patients, adrenal axis in 48.6.% and thyroid axis in 40.5%. Most patients have increased levels of serum PRL[8]. Our patient had panhipopituitary and hyperprolactinemia. The diagnosis of a intra sellar aneurysm is based in the CT scan, MRI and digital cerebral angiography. In our patient real diagnosis was only found in the histopathology biopsy, when a aneurysm wall and thrombosis component were found after a minimally invasive transseptal endoscopic surgery[13].

The treatment of cavernous intra and parasellar aneurysm is based in direct and indirect surgical methods. Direct options are more difficult and may have a higher risk of morbidity for patients with aneurysms arising from the cavernous internal carotid segment. This treatment includes neck clipping, direct obliteration with trapping and sacrifice of the internal carotid artery, aneurysm-wall reinforcement, and excision of the aneurysm with reconstruction of internal carotid artery[5]. Indirect repair consists of either proximal occlusion of the internal carotid artery or trapping of the aneurysm, with isolation proximally and distally[5]. The minimally invasive technique of endovascular neuroradiology has added more options to the therapeutic armamentarium of this pathology.

A thrombosed intra and parasellar aneurysm is an important differential diagnosis of apparent intrasellar tumor with lateral extension, even if hypopituitarysm and hyperprolactinemia exist. MRI and digital angiography are more reliable than CT for detecting the aneurysm. Hypopituitarism is usually irreversible, and permanent hormone replacement therapy is necessary.

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