A Bi-Frontal Ischemic Stroke Revealing a Unilateral Pheochromocytoma

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DOI: 10.36347/sasjm.2021.v07i02.001 | Received: 26.01.2021 | Accepted: 15.02.2021 | Published: 18.02.2021

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Abstract

Pheochromocytoma is an infrequent and severe disease; it occurs early, its main manifestation is hypertension and its association with signs of seriousness makes it severe. Our observation is an example of this; the pheochromocytoma is revealed by a bi-frontal ischemic stroke which dominated the initial picture in a 51-year-old adult. Imaging guided the diagnosis (cerebral CT, ultrasound and abdominal CT), confirmed by the determination of urinary catecholamines.

Keywords: Pheochromocytoma, bi-frontal ischemic stroke, arterial hypertension, intensive care.

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INTRODUCTION

Pheochromocytoma is a rare tumor of the adrenal medulla, secreting catecholamines. It is often revealed by paroxysmal or permanent hypertension [1]. Confronted with the problem of diagnosis, given its rarity and the various associated clinical signs, the diagnosis is based on the dosages of urinary catecholamines, their precursors or their metabolites.

Imaging, in particular MRI and meta-iodobenzylguanidine (MIBG) scintigraphy, provide valuable diagnostic assistance. Therapeutic care is multidisciplinary. The diagnostic confirmation is based on the pathological study of the surgical specimen.

We report the case of a 51-year-old adult who presented with a unilateral pheochromocytoma revealed by a bi-frontal ischemic stroke (stroke) admitted to intensive care.

PATIENT AND OBSERVATION

51-year-old patient, with a history of hypertension poorly monitored for 8 years, the onset of symptoms dates back to one day of his admission to intensive care with the installation of paroxysmal hypertensive peaks, associated with profuse sweating, and palpitations.

On admission to intensive care, the patient is comatose with a GCS at 08/15 with a right flaccid hemiplegia and left hemiparesis, the initial brain scan was normal.

The examination found a comatose patient, afebrile, with a weight of 78kg for a height of 180cm, a flexible neck and the Signs of Koernig and Brudzinski are negative, the lumbar puncture brings back clear fluid with less than 03 white elements / mm\textsuperscript{3}, normal glycorachia and proteinorachia sterile culture after 48 hours, the evolution is marked by the onset of status epilepticus, and the patient is placed on artificial ventilation with antihypertensive treatment with anticalcics (amlodipine 10 mg / 24h) and beta-blocker (and antiepileptic: phenobarbital, midazolam).

The control brain scan 48 hours after admission reveals deep bilateral parietal areas of hypodense. Angio MRI revealed ischemic stroke of the anterior cerebral artery territory with distal right and left thrombi and a thin aspect of the anterior communicating artery.
The EEG was normal

ECG: was of regular sinus rhythm with diastolic-type HVG with repolarization disorder. Transthoracic echocardiography was normal. Ultrasound of the supraortic trunks revealed atheromatous plaques without significant stenosis. The evolution was marked by blood pressure instability with persistence of hypertensive peaks; the patient was then placed on alpha blockers and anticalcics.

The assessment of the clinical confirmed arterial hypertension was supplemented by an abdominal scan which objectified the presence of a right adrenal tissue lesion which is discreetly enhanced with the injection of contrast product, measuring 21/15 mm without signs of extension. In addition, the abdominal scan revealed microlithiasis of the kidneys with a small left kidney and a large right kidney. A pheochromocytoma is strongly suspected.

The test of urinary metanephrines on 24-hour urine shows an elevated normetanephrine at 0.96 mg / 24 h (N 0.07 to 0.46). The management was medical by calcium blockers and beta blockers for a week, after his release from the neurology department he was scheduled for surgical treatment of the tumor.

DISCUSSION

Pheochromocytoma is a rare tumor of the adrenal medulla, secreting catecholamines. It is often revealed by paroxysmal or permanent hypertension [1]. Pheochromocytoma is a tumor that develops at the expense of chromaffin cells in the adrenal medulla or extra adrenals embryonic chromaffin residues producing catecholamines. Pathology often found in adults, and rarely seen in children [2].

DIAGNOSIS

The diagnosis should be made as early as possible, given the severe complications that can appear, including ischemic stroke [3]. Clinical polymorphism indicates adrenergic hypersecretion or tumor growth. The picture is dominated by Ménard's clinical triad: pulsatile headache, palpitations and sweating [4]. As for arterial hypertension, it is systolodiastolic, most often permanent, and tends to be less severe than in children [5].

Hemiplegia has brachiofacial predominance or is proportional depending on the cortical or central predominance of the ischemic impact. The disturbances of consciousness are moderate or absent except in massive infarctions or giving rise to prolonged convulsions [6].

Biology allows the diagnosis by the demonstration in excessive quantity during urinary and blood dosage of free catecholamines (adrenaline, noradrenaline, and dopamine) and their metabolites (vanillylmandelic acid, methoxylated derivatives) [7].

The most reliable measurement at present is the urinary methoxide dosage with a sensitivity of over 99%. Imaging is important for confirming the diagnosis, assessing the extent of ischemic parenchymal lesions and their degree of necrosis, the mechanism of arterial ischemia, and determining the location of the tumor prior to surgical treatment.

The chest x-ray looks for a possible chest location. Abdominal ultrasound is the screening examination to locate an isolated pheochromocytoma. The exams of choice today are computed tomography and magnetic resonance imaging. Meta-iodobenzylguanidine (MIBG) scintigraphy allows whole-body mapping, for the detection of multiple tumors or ectopic locations [6].

MECHANISM

The cause is either a low cerebral flow with arterial vasospasm, or a hypertensive mechanism caused by adrenergic hypersecretion, or the migration of emboli. Acute hemiplegia is the usual mode of revelation of ischemic cerebrovascular accidents in children [6]. A review of the literature revealed 25% of perioperative deaths in relation to blood pressure variations such as hyper or hypotension [8].

Therapeutic care

First, a preparation must be made, the objectives of which were specified by Roizen et al. in 1978 [9]: "to normalize the arterial pressure and the electrocardiographic repercussions of the underlying cardiopathies most often hypertrophic, while bringing the patient on the side of the side effects, orthostatic hypotension and nasal congestion".
The modalities differ according to the teams [10]. The use of drugs that block alpha adrenergic receptors aims to correct hypertension and prevent paroxysmal outbreaks. These alpha blockers also make it possible to normalize blood volume when it is reduced, and to re-sensitize the alpha receptors [10], they have profoundly modified the operative prognosis of pheochromocytoma.

Calcium blockers from the dihydropyridine family have been shown to be effective in preparing pheochromocytomas for surgery [11, 12]. They have few side effects.

The goal of treatment is to completely remove the tumor, which is the source of the adrenergic hypersecretion. This surgical treatment must be preceded by a preoperative preparation lasting 7 to 14 days, in order to lower and stabilize the blood pressure figures to prevent any accident of intraoperative release of catecholamines, while avoiding an excessive lowering, damaging to the myocardial and cerebral tissue.

Correction of blood volume within 18 hours of surgery is essential [13]. The surgical treatment, strictly speaking, consists of a complete excision of the pheochromocytoma by conventional or laparoscopic surgery [14]. The rule of thumb for this surgery is to “gently dissect the patient from the tumor, not the patient's tumor.” Elevation ligation of the adrenal veins prior to all tumor traction and pressure maneuvers is essential [15].

From an anatomopathological point of view, pheochromocytomas are characterized by an important architectural polymorphism from one area to another. They are made up of pheochromocytes organized in trabecular or banded formations. The histological diagnosis is supported by the demonstration, in the tumor cells, of argyrophilic granulations by Grimelius staining and by immunohistochemical techniques [16]. Histological analysis of the tumor does not make it possible to distinguish a benign form of a malignant form. According to the classification of the World Health Organization (WHO), the malignancy of these tumors is defined by the presence of metastases at an extra-lymph node site (lymph nodes, bone, liver) [17].

The most widely used malignancy score is the Pheochromocytoma of the Adrenal Gland Scaled Score (PASS), by Thompson [18]. A score less than 4 are in favor of benignity, greater than 6, of malignancy [19].

EVOLUTION

The evolution of untreated pheochromocytoma is always serious, often fatal in the case of pheochromocytomas operated on. In the short term, their prognosis is now very good in the majority of cases, perioperative mortality having decreased considerably [16]. The difficulties are mainly linked to the diagnosis of recurrence; in fact, a recurrence can occur many years after the initial diagnosis, the differences ranging from 1 to more than 20 years in adults [16].

CONCLUSION

Pheochromocytoma is an infrequent and severe pathology; it occurs at an early age, the clinical presentation is polymorphic and is dominated by Ménard's triad in adults.

A stroke is a terrible complication, radiological and biological investigations are very important tools in the etiological diagnosis before surgical treatment.

Conflicts of interest

All authors declare no conflict of interest.

The contributions of the authors

All the authors participated in the management of the patient; all the authors have read and approved the latest version of the manuscript.

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