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CASE REPORT

Acute type A aortic dissection in a patient with paraganglioma☆

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KEYWORDS
Aortic dissection;
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Alpha- and beta-blockers

Abstract Acute aortic dissection is the most common acute aortic syndrome. It is more prevalent in males and in the elderly, and has a high mortality. Hypertension is the main risk factor. Diagnosis is based on clinical features, laboratory tests and imaging exams. Treatment is usually surgical, although in some cases an endovascular approach is an alternative.

Paraganglioma is an uncommon neuroendocrine tumor. Most produce catecholamines, and so usually manifest with hypertensive crisis, palpitations, headache and sweating. This tumor is diagnosed by measurement of plasma or urinary catecholamines and by computed tomography, magnetic resonance imaging and ^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy. Surgery is the only potentially curative treatment.

This case report describes a female patient with type A aortic dissection associated with paraganglioma. This association is very uncommon and the management of both conditions presents a challenge.

PALAVRAS-CHAVE
Dissecção aórtica;
Paraganglioma;
Hipertensão arterial;
Catecolaminas;
Tratamento cirúrgico;
α e β-bloqueantes

Dissecção aórtica aguda do tipo A em doente com paraganglioma

Resumo A dissecção aórtica aguda é a síndrome aórtica aguda mais frequente, ocorrendo predominantemente no sexo masculino e em idosos, estando associada a uma elevada mortalidade. Existem vários fatores de risco, destacando a hipertensão arterial. O diagnóstico é feito com base na clínica, exames laboratoriais e imagiológicos. A terapêutica habitual baseia-se na abordagem cirúrgica, existindo em alguns casos a alternativa do tratamento endovascular.

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O paraganglioma é um tumor neuroendócrino raro. A maioria produz catecolaminas e manifesta-se frequentemente por crises hipertensivas, palpitações, cefaleias e hipersudorese. O diagnóstico deste tumor passa pelo doseamento de catecolaminas urinárias e séricas, e pela realização de TC toracoabdominopélvica, ressonância magnética ou cintigrafia com ^{123}MIBG. O tratamento cirúrgico é o único tratamento potencialmente curativo. Neste artigo, descreve-se um caso clínico de uma doente com uma dissecção aórtica do tipo A, associada a um paraganglioma. Esta associação é extremamente rara e a abordagem de ambas as patologias constitui um desafio.

Introduction

Acute aortic dissection and paraganglioma are both rare conditions. Acute aortic dissection is defined as an intimal tear with passage of blood from the aortic lumen into the media, forming a false lumen.[1] Paragangliomas are tumors that originate in extra-adrenal chromaffin cells and may or may not produce catecholamines[2]; they are one of the secondary causes of hypertension, which is a risk factor for aortic dissection.

We present the case of a female patient with type A aortic dissection and paraganglioma, and highlight the rarity of this association.

Case report

A 49-year-old female patient, black, born in Africa but resident in Portugal for 12 years, had a history of hypertension and hyperthyroidism. On May 20, 2010, she suffered headache, self-medicated with nebivolol and bioflavonoids. She then she suffered chest pain with no relieving factors and went to the emergency department of another hospital, where she reported two episodes of vomiting and two of hemoptysis. On observation she was alert, cooperative and oriented in time and space, hypertensive (blood pressure 200/140 mmHg) and with a heart rate of 87 bpm. There were no other abnormalities on physical examination.

Laboratory tests showed hypochromic microcytic anemia, leukocytosis (leukocytes 16 200 Ul), thrombocytopenia (platelets 141×10^9/l), elevated D-dimers (8.01 μg/ml), low-density lipoprotein (1032 U/l), aspartate aminotransferase (60 U/l) and C-reactive protein (2.4 mg/dl), and negative troponin I (0.016 ng/ml).

The electrocardiogram (ECG) (Figure 1) showed no significant alterations but the chest X-ray (Figure 2) revealed increased pulmonary vascularity and enlarged cardiac silhouette and mediastinum.

These findings prompted an urgent thoracic-abdominal-pelvic computed tomography (CT) angiogram, which showed an aneurysmal aortic dissection that extended from the aortic root to the right common iliac artery; densification of mediastinal fat with no signs of recent hemorrhage; a heterogeneous solid nodular formation around 45 mm in diameter on the right side of the aorta at the level of the emergence of the celiac trunk and in close contact with the inferior vena cava, displacing the hepatic artery anteriorly and displacing and compressing the inferior vena cava laterally; and dilatation of the proximal segment of the inferior vena cava, with a heterogeneous lumen up to the convergence of the renal veins, probably due to thrombosis (Figure 3).

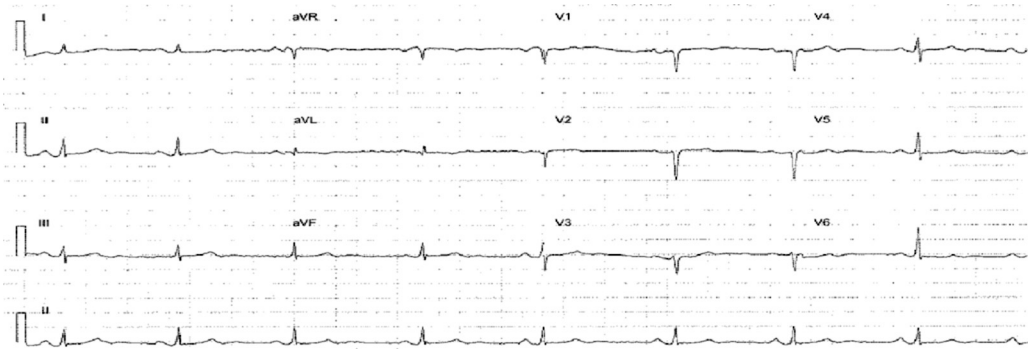

Figure 1 Electrocardiogram.