

Coronary aneurysms in a child: an unusual presentation of pseudovasculitis

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ABSTRACT

Abnormalities of the coronary arteries in children are rare and Kawasaki disease is the most common cause of acquired coronary disease in a paediatric population. We report a case of a female child with coronary artery aneurysms and convulsions, who was diagnosed with Kawasaki disease. Due to systemic arterial hypertension and persistence of high inflammatory markers after treatment with high dose glucocorticoid and intravenous immunoglobulin, further investigation was performed and revealed a pheochromocytoma. Surgical removal led to normalisation of blood pressure and laboratory parameters. Periodic echocardiography studies revealed progressive reduction of coronary aneurysms, with complete normalisation after 8 months. This is the first case described of coronary aneurysms presenting as a pseudovasculitis syndrome associated with pheochromocytoma.

Case report

An 8-year-old Caucasian girl who was previously healthy presented in the emergency room (ER) with generalised tonic-clonic convulsions. She complained of blurred vision and headache in the last three days. Two months before, she had diffuse erythema and fever, but she was afebrile at ER admission. Her fundoscopy showed some haemorrhage spots. She underwent magnetic resonance imaging (MRI), which showed alterations compatible with central nervous system vasculitis; haemorrhage and tumours were excluded (Fig. 1A). Pulse therapy with methylprednisolone was initiated, and the convulsions stopped. Echocardiography revealed right and left coronary artery aneurysms (Fig. 1B), and a diagnosis of Kawasaki disease was suspected. She received intravenous immunoglobulin (2g/kg) and was referred to our department. Her physical examination revealed livedo reticularis, papilloedema and systemic arterial hypertension (230 x 140 mmHg). Laboratory evaluation showed high erythrocyte sedimentation rate (108 mm/1st hour), high C-reactive protein (129 mcg/L), hypoalbuminaemia (2.6 g/dL), and

hypergammaglobulinaemia (2.1 g/dL). Renal parameters were normal: blood urea nitrogen (7.9 mg/dl), creatinine (0.32 mg/dl) and urinalysis (absence of proteinuria, haematuria or urinary casts). She had no response to anti-hypertensive therapy and persistence of high inflammatory markers, even using a high dose of glucocorticoid for 2 weeks. Due to this clinical picture, a thoraco-abdominal angiographic computed tomography was performed and demonstrated an adrenal tumour on the left side (Fig. 2), raising suspicion of a pheochromocytoma. Urinary norepinephrine (300 mcg/24h; normal range: 14–80 mcg/24 h) and metanephrine (76.67 mg/24h; normal range: 2–12 mcg/24 h) were elevated and confirmed the pheochromocytoma diagnosis. Her left adrenal was removed, and she showed a disappearance of livedo reticularis and normalisation of blood pressure after the surgery. Laboratory tests were performed a month after the surgical procedure and demonstrated an impressive improvement in the previous abnormalities (erythrocyte sedimentation rate: 9 mm/1st hour, C-reactive protein: 3 mcg/L, albuminaemia: 3.6 g/dL, and gammaglobulinaemia: 1.2 g/dL). After six months, a new central nervous system MRI was normal, and periodic echocardiography studies revealed progressive reduction of coronary aneurysms, with complete normalisation after 8 months.

Discussion

Abnormalities of the coronary arteries in children are very rare and can be congenital or acquired. Congenital coronary aneurysms occur isolated or accompanying other heart defects and patients usually present with cardiovascular and respiratory symptoms, while inflammatory features are absent (1). The most common cause of acquired coronary artery anomalies in a paediatric population is Kawasaki disease (KD) (1).

Kawasaki disease is a childhood vasculitis. Its incidence varies throughout the world, with higher rates in Japan (approximately 112 cases per 100.000 children <5 years old) (2). The most worrisome complication is the devel-

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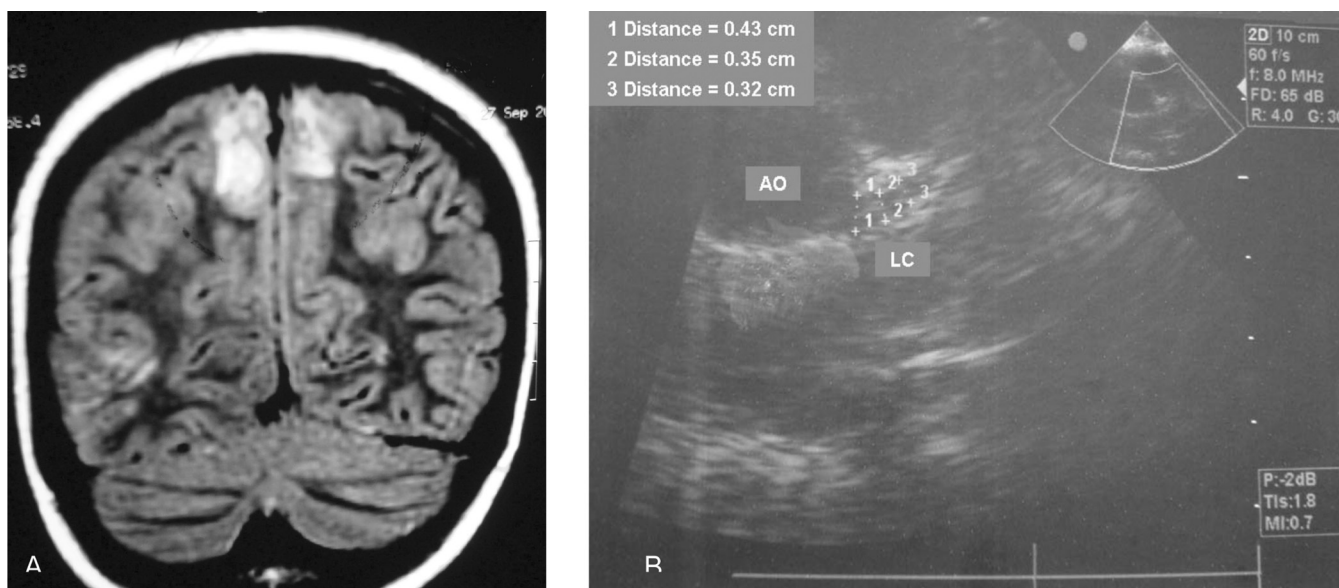


Fig. 1. Images suggestive of vasculitis: Magnetic resonance imaging (T2) showing lesions compatible with central nervous system vasculitis [A] and Echocardiography revealing an aneurysm of the proximal portion of the left coronary (LC) artery [B].

opment of coronary artery aneurysms (3-4), which are present in 15 to 25% of untreated children and may lead to ischaemic heart disease and sudden death. Prompt treatment during the first 10 days of illness considerably reduces the risk of this complication (2).

Although this patient did not fulfil the criteria for KD (5), the detection of a coronary aneurysm upon echocardiography and the previous history of fever and exanthema associated with high levels of inflammatory parameters strongly suggested a diagnosis of KD. Also, neurological manifestations such as convulsions may be present in KD patients in a frequency that varies from 1.1 to 3.7% and seems to be more frequent in patients with more severe disease (6-7).

The lack of response to intravenous immunoglobulin and methylprednisolone pulses, the maintenance of elevated inflammatory markers, associated to the presence of severe and refractory systemic arterial hypertension presented herein raised suspicions of an alternative diagnosis.

In the general population the most common cause of coronary aneurysms is atherosclerosis, but this is particularly important in adults (8). Coronary aneurysms have been described as complication of inflammatory vascular diseases such as polyarteritis nodosa, syphilis, systemic lupus erythematosus, Tak-

Fig. 2. Abdominal computed tomography demonstrating a hypervascularised solid tumour on left adrenal measuring 1.6 x 1.3 cm in the patient with pheochromocytoma.



yasu arteritis, Behçet's disease, Wegener's granulomatosis, microscopic polyangiitis and Churg-Strauss syndrome (9-13). Coronary aneurysms can also follow connective tissue disorders (Ehlers-Danlos and Marfan's syndrome) and other pathologies like candidosis, primary hyperaldosteronism, and following coronary artery revascularisation procedures (balloon angioplasty, laser, atherectomy) and chest trauma (9).

In our patient, due to the refractory systemic arterial hypertension, we performed a thoraco-abdominal angiographic computed tomography, which demonstrated an adrenal tumour. The finding of elevated urinary norepinephrine and metanephrine led to diagnosis of pheochromocytoma. Interestingly, there are a few cases of pheochromocytoma manifesting as pseudovasculitis syndromes, but, to

our knowledge, there is no previous report of coronary aneurysms induced by pheochromocytoma.

Similar to the case presented here, a previous report showed a misdiagnosis of central nervous system vasculitis by angiographic imaging in a patient who had a pheochromocytoma (14). Another case of pseudovasculitis associated with pheochromocytoma characterised by heliotrope rash, livedo reticularis, focal infarcts of the fingertips, arthritis, severe hypertension, pulmonary and cardiac failure, and high erythrocyte sedimentation rate was described (51). There are also reports of intestinal arteritis (16), cutaneous vasculitis (17) and cryoglobulinemia (18) associated with pheochromocytoma.

Pheochromocytoma is a rare clinical entity in children and may present with a variety of symptoms, including headache, sweating, flushing, palpitations, blurred vision, syncope, tremor, and weight loss. Complications described are cardiomyopathy, severe hypertensive crisis, stroke, convulsions mimicking vasculitis, mass effect and multi-organ failure (19).

The authors conclude that pheochromocytoma should be taken into account in cases of suspected systemic vasculitis in which hypertension is a dominant finding and response to standard treatment is poor.

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