

Fibromuscular dysplasia of the brachial artery associated with unilateral clubbing

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Abstract

A 46-year old male patient was admitted with a history of an extremely painful right upper arm, associated with unilateral clubbing. Duplex scanning and magnetic resonance imaging were suggestive of a pseudo-aneurysm of the brachial artery. Digital angiography showed an irregular brachial artery, associated with a small pseudo-aneurysm. The brachial artery was partially resected and reconstructed with a venous interposition graft. Pathological examination provided the final diagnosis of fibromuscular dysplasia. Although more encountered in women, this case report describes the occurrence of fibromuscular dysplasia in an unusual location in a male patient with a long-term follow-up.

Keywords: Fibromuscular dysplasia • Brachial artery • Clubbing

CASE REPORT

A 46-year old man was referred because of a 13-year history of pain in the right upper arm, aggravated after a whiplash. This pain was localized at the medial part of the upper arm and radiated to the shoulder and forearm. Due to this irradiating pain, there was severe functional limitation. Unilateral clubbing of the fingers of the right hand was found during physical examination. In 1995, the patient had undergone a surgical exploration of the arm at another institution without any improvement. Duplex scanning showed a vascular pseudo-aneurysmal malformation of the right brachial artery with focal widening. Magnetic resonance angiography confirmed the previous finding, demonstrating a proximal small aneurysm of the right brachial artery with secondary impingement on the median nerve. No mass or neurinoma were detected. Digital angiography of the right arm showed an irregular mid-part of the brachial artery, associated with a small pseudo-aneurysm as a possible consequence of arteritis (Fig. 1). Peripheral emboli could not be documented. Preoperative differential diagnosis was made between cervicgia, neurological problems and muscle problems and these were excluded. Immunological evaluation did not provide any evidence of systemic disease. Because of the severe complaints and a non-functional right arm, a surgical re-exploration of the brachial artery was performed. The diseased part of the right brachial artery was resected and a venous interposition graft was constructed. Neurolysis of the median nerve was done as well. Definitive pathological analysis showed fibromuscular dysplasia (FMD) with mostly intimal changes of the right brachial artery (Fig. 2). Postoperative recovery was uneventful. Further investigation of

the renal and carotid arteries by magnetic resonance angiography showed no signs of fibromuscular dysplasia.

At 6-year follow-up, clubbing of the right hand and the subjective symptoms of pain and cold sensations in the right upper limb had disappeared. Duplex ultrasound showed a triphasic signal with good patency of the venous bypass. No sign of stenosis was encountered.

DISCUSSION

Fibromuscular dysplasia (FMD), first described by Leadbetter and Burkland in 1938, is a non-atherosclerotic non-inflammatory angiopathy that causes narrowing of medium-sized arteries, characterized by fibrodysplastic changes. With a frequency lower as 1%, FMD most commonly affects the renal arteries, reported in 60–75% of the cases. Cerebrovascular involvement is present in 25–30%. Uncommon reported sites are visceral, upper and lower limb arteries, aorta and coronary or pulmonary arteries. In 28% of patients, more than one arterial region is affected. Although FMD has been associated with genetic and hormonal anomalies, coagulation disorders, stress and smoking, the true cause of FMD remains unknown. The disease has a preference for young to middle-aged women. No particular symptoms are pathognomonic for FMD. Patients with FMD of the brachial artery may primarily present with ischaemia and/or nerve compression-related symptoms. FMD has been classified according to the arterial wall layer that is predominantly affected: intimal, medial and adventitial fibroplasias. Medial fibroplasia accounts for 90% of all cases. In our patient, the intima was primarily affected [1, 2].

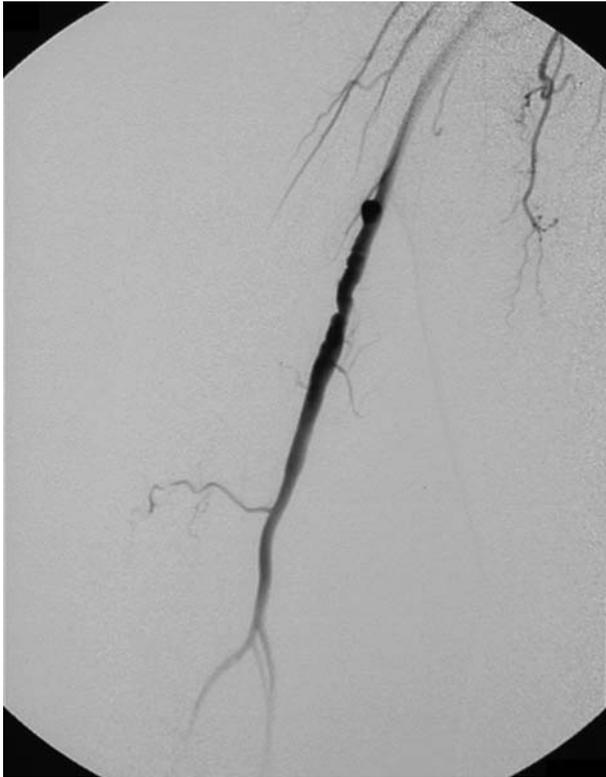


Figure 1: Angiographic image of fibromuscular dysplasia of the right brachial artery.

Clubbing of digits can be explained by hypoxaemia due to disturbances in blood flow in the brachial artery. Treatment of FMD of the brachial artery may consist of percutaneous transluminal angioplasty (PTA) by placing a stent or an open procedure. There is no proof that PTA is superior to the surgical procedure, although PTA is more indicated for short segment stenosis.

FMD of the brachial artery is rare and has only been reported in 21 cases in the last 20 years [3, 4]. A clear review of these patients is given by Rice *et al* [3]. As most of them comprised females, our case is unique in representing unilateral clubbing at an uncommon site in a male patient with a long-term follow-up period.

Conflict of interest: none declared.



Figure 2: Section with hematoxylin-eosin stain of brachial artery with marked intimal hyperplasia without signs of atherosclerosis or inflammation. Mucoid degeneration of tunica media and focal narrowing of lumen were observed.

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