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Bilateral Internal Carotid Artery Fibromuscular Dysplasia Resulting in Ischemic Stroke in an Elderly Woman

González Fabio [1,*], MD, Saucedo Miguel Angel [1], MD, Bala Marta Inés[1], MD, Uribe Roca Claudia[1], MD, León Cejas Luciana[1], MD, Fernández Pardal Manuel María[1], MD, Reisin Ricardo[1] , MD, Bonardo Pablo[1], PhD, MD

[1] Neurology department - Hospital Británico de Buenos Aires

Abstract

Background— There is limited data regarding fibromuscular dysplasia in elderly patients particularly those with ischemic stroke.

Case Description—An 84-year-old woman was admitted because she had sudden onset episodes of involuntary movement her left hand compatible with alien hand syndrome. On physical examination, she was found to have sensitive agnosia and diminished deep sensory perception of the left arm. Brain magnetic resonance imaging demonstrated an area of restricted diffusion in right post Rolandic area. Computed tomographic angiography revealed classical "string of beads" in the middle and distal cervical portion of both internal carotid arteries suggestive of fibromuscular dysplasia.

Conclusion— We report a late presentation of fibromuscular dysplasia associated with ischemic stroke in an elderly woman highlighting the need for recognition of the disease in elderly ischemic stroke patients.

Keywords— Fibromuscular dysplasia, Internal carotid artery, Ischemic stroke, Elderly.

INTRODUCTION

Fibromuscular dysplasia is an infrequent non atherosclerotic, non-inflammatory artery disease that usually affects large and medium-sized arteries. Prevalence in the general population is unknown, but indirect estimates suggest that approximately 4% of persons may be affected. Fibromuscular dysplasia affects more frequently young Caucasian women, but has been rarely reported in the very elderly population.[1,2] Here we describe an elderly woman with an atypical stroke presentation secondary to fibromuscular dysplasia.[3]

CASE DESCRIPTION

An 84-year-old woman with a history of hypertension, dyslipidaemia and without previous diagnosis of vascular disease was admitted to our hospital because she had sudden onset episodes of involuntary movement her left hand. On physical examination, she was found to have sensitive agnosia and diminished deep sensory perception of the left arm. The laboratory tests were unremarkable. Brain magnetic resonance imaging demonstrated an area of restricted diffusion in right post Rolandic area. Based on the clinical presentation and imaging data, we diagnosed Allien Hand syndrome involving the left upper extremity. Computed tomographic angiography revealed classical "string of beads" appearance in the middle and distal cervical portion of both internal carotid arteries suggestive of fibromuscular dysplasia (Figure 1). Transoesophageal echocardiogram and Holter monitoring did not identify any cardiac embolic source. No involvement of other arteries was seen. She was treated with aspirin 100 mg and rosuvastatin 20mg daily. The patient did not experience any stroke recurrence during 24 months of follow up.

DISCUSSION

Our patient presented with abnormal movements on her left arm compatible with “alien hand syndrome” at the age of 84 years. Further investigations demonstrated an ischemic stroke in right post Rolandic region with fibromuscular dysplasia of internal carotid arteries. Fibromuscular dysplasia is characterized by the proliferation of connective tissue and muscle fibres within the arterial vessel walls; causing irregular thickening through proliferation of smooth muscle and fibrous tissue within the media, which narrows the lumen.[4,5] It is classified into 3 categories according to the vessel wall layer that is affected: intimal (intimal fibroplasia), medial (medial dysplasia, peri medial fibroplasia and medial hyperplasia) and the adventitia (adventitial fibroplasia). The

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***Corresponding Author:** Gonzalez Fabio, Servicio de Neurología – Hospital Británico de Buenos Aires, Ciudad Autónoma de Buenos Aires - Argentina, e –mail: fabiogonzalezclinicas@yahoo.com, phone: +5491143041081.

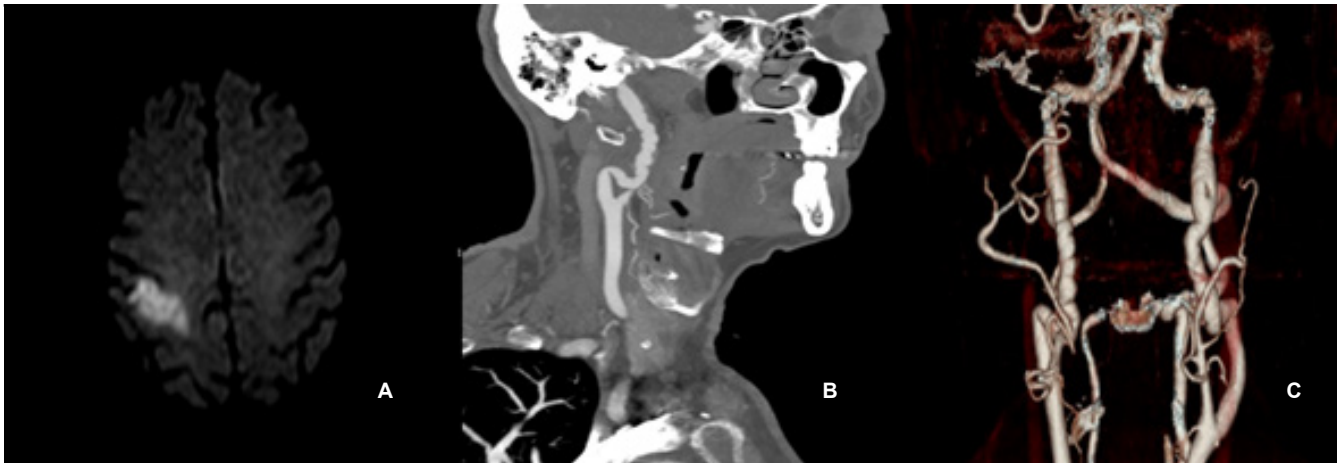

FIGURE 1: A) Brain magnetic resonance imaging (diffusion weighted imaging): Brain magnetic resonance imaging demonstrated an area of restricted diffusion in right post Rolandic area, B) and C) Computed tomographic angiography demonstrates the typical "pearl neck lace" pattern consistent with fibromuscular dysplasia.

most common type is medial dysplasia (70%).[5] Fibromuscular dysplasia generally involves renal arteries (60–75%), cervicocranial arteries (25–30%), visceral arteries (9%), the arteries of the extremities (5%), and rarely also coronary arteries.[6] Fibromuscular dysplasia results in bilateral involvement in 50% of the patients.[6,2] Infrequently, aneurysms in the cerebral circulation may be seen.[4,5] Annual rates of ischemic strokes secondary to fibromuscular dysplasia range between 0.75% and 4.9%.[6] Approximately 20% of all ischemic strokes in patients younger than 45 years may be related to fibromuscular dysplasia.[7] In an international registry of 447 patients, carotid and vertebral arteries were affected in 74% and 36% of the patients with fibromuscular dysplasia, respectively. Transient ischemic attacks were observed in 13.4% and 10% of the patients, respectively.[1] The disease is very infrequent in elderly patients. Bagh et al.[8] reported that only 16.7% of 1016 patients with fibromuscular dysplasia were aged 65 years or greater. Elderly patients were predominantly asymptomatic with a low occurrence of ischemic stroke.

Our case highlights that fibromuscular dysplasia as an etiology for ischemic stroke even in elderly patients.

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