Case report

Livedo reticularis in multiple system atrophy: case report

Livedo reticular em atrofia de múltiplos sistemas: relato de caso

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INTRODUCTION

Multiple system atrophy (MSA) is a neurodegenerative disorder characterized by autonomic failure in combination with parkinsonism or cerebellar ataxia, and pyramidal signs. Patients can be classified depending if parkinsonism (P) or cerebellar (C) signs are more prominent (AMS-P or AMS-C).[1,2] Manifestations of autonomic failure involving cutaneous system are rare and include acrocyanosis, erythromelalgia and Raynaud phenomenon. Livedo is a physical finding consisting of macular, violaceous, connecting rings that form a netlike patterned erythema of the skin. There are two types of livedo: reticularis (LR) and racemosa. The first one is characterized by diffuse and homogeneous and the last by irregular with broken circular segments, and persistence on warming. In neurology, the reticularis and racemosa presentations could be related to: amantadine (reticularis possibly related to cutaneous NMDA receptors), Sneddnon's syndrome (racemosa with stroke), Divry-van Bogaert disease (racemosa, seizures, pseudobulbar symptoms, dementia, pyramidal-extrapyramidal signs, related to stroke, white matter disease and cerebromeningeal angiomatosis), cerebral thromboangiitis obliterans (racemosa with clinical picture similar to Divry-van Bogaert, due to multiple stoke and white matter disease, without cerebromeningeal angiomatosis), antiphospholipid syndrome (antiphospholipid antibodies with livedo racemosa eventually related to multiple neurologic presentations, such as stroke, chorea, migraine, dementia, and seizures), neurolupus (racemosa or reticularis), polyarteritis nodosa (rare cases with racemosa and stroke or polyneuropathy), migraine (general livedo racemosa, this association increasing the risk of stroke), cholesterol embolization syndrome (distal showering of cholesterol crystals eventually leading to livedo racemosa and stroke).[3] The pathophysiology of LR consists in increased visibility of the cutaneous venous plexus caused by venodilation or deoxygenating of blood. Examples of potential causes of venodilation include altered autonomic nervous system function, circulating venodilators and/or local hypoxia. Deoxygenating is mainly caused by decreased cutaneous perfusion, which can be caused by decreased arteriolar inflow or increased resistance to venous outflow.[4]

We describe a case of MSA-P in which LR was present on physical examination.

CASE REPORT

Man, 76-year-old, initiated two years ago with rigidity, bradykinesia, early and frequent falls, progressing to wheelchair within one year. Besides that, he presented severe dysphagia and dysarthria. His first

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diagnosis was Amyotrophic Lateral Sclerosis. Physic examination revealed global rigidity, more severe in lower limbs, bradykinesia, hyperreflexia with bilateral Babinski sign, and dysarthria. LR was observed in both thighs, independent of cold exposure (Figure 1). He was not in use of any drug that could lead to LR. Tilt test revealed autonomic failure and brain Magnetic Resonance Imaging (MRI) showed severe cerebellar and pons atrophy. There was no response to levodopa.

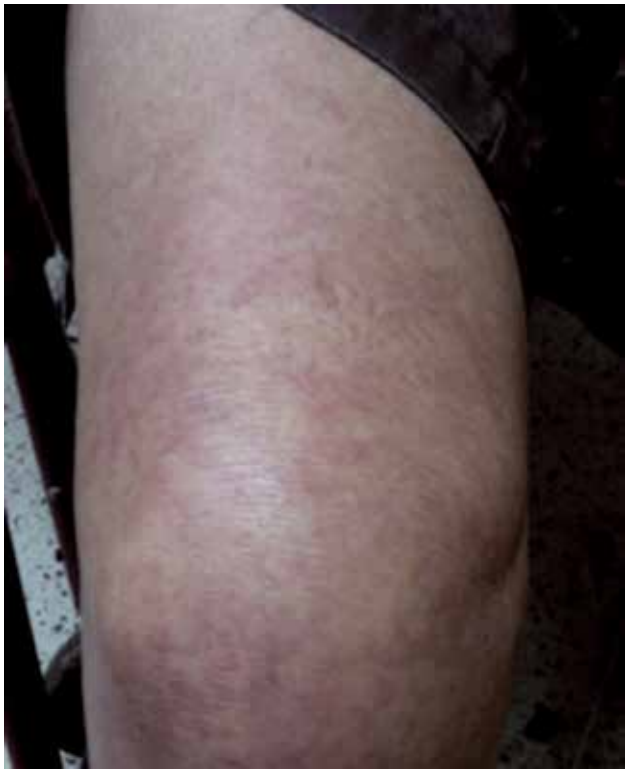

Figure 1. Livedo reticularis independent of cold exposure.

DISCUSSION

MSA probably is an underdiagnosed disorder, generally affects patients over 50 years of age, being more common in men.[5] Patients with MSA can initially present parkinsonism, undistinguishable from Parkinson disease (PD), or cerebellar signs, undistinguishable from sporadic ataxia. The correct diagnoses in early stages may be difficult to be established. Other indicators for diagnosis ("red flags") are: symmetric signs, rapid progression, absence of tremor, lack of response to L-dopa, and early autonomic dysfunction.[1] Even though dysautonomia can occur in both, MSA and PD, the presence of severe and early autonomic failure strongly suggests MSA.

Despite the fact that LR can be a side effect of some drugs, such as amantadine for example, there are few scientific reports about this phenomenon related to AMS. This cutaneous manifestation is more common in women and develops in up to 40% of patients taking amantadine, and progression to ulceration is possible with continued use.[3] Ulcers typically resolve quickly with discontinuation, but resolution of the LR may take longer time. Patients with parkinsonism and LR, the first diagnosis to be consider is relation with amantadine, and if not present, manifestation of dysautonomia could be related to it. In the present case there were symmetric signs, no response to L-dopa and dysautonomia, which represent "red flags" for MSA. Moreover, cerebellar and pons atrophy on MRI corroborated to elucidate the case.

We have not found any report in the literature of LR as a manifestation of autonomic dysfunction in MSA patients. This cutaneous manifestation associated with parkinsonian syndrome, without history of drugs that could lead to LR, should be considered as an autonomic failure, and it is an important clue that suggests MSA as the underlying disease.

Conflict of interest

There is no conflict of interest to declare.

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