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(CASE REPORT)

SNEDDON SYNDROME REVEALED BY ISCHEMIC STROKE AND LIVEDO RACEMOSA IN A 69-YEAR-OLD MAN: A CASE REPORT

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ABSTRACT

Background: Sneddon syndrome is a rare, non-inflammatory thrombotic vasculopathy of small- and medium-sized arteries, defined by the association of livedo racemosa and cerebrovascular events. It classically affects young to middle-aged women, which makes recognition in elderly patients particularly challenging and easily overlooked.

Case Presentation: We report the case of a 69-year-old man with a history of hypertension and diabetes, admitted to the neurology department for an acute ischemic stroke presenting as right-sided hemiplegia and aphasia. Physical examination revealed a widespread livedo racemosa of the trunk and lower limbs, which prompted a dermatological evaluation and led to the diagnosis of Sneddon syndrome. Brain computed tomography showed a left fronto cortico-subcortical hypodensity in the territory of the left middle cerebral artery, consistent with a subacute ischemic infarction. The antiphospholipid antibody work-up was negative, defining an antiphospholipid-antibody-negative form of the syndrome. A skin biopsy was not performed, as the eruption was clinically typical of livedo racemosa. The patient was managed with antiplatelet therapy and optimization of vascular risk factors, but his condition deteriorated during hospitalization and he died from cardiogenic shock.

Conclusion: This case highlights that Sneddon syndrome should remain in the differential diagnosis of stroke with cutaneous livedo even in advanced age, and that a simple skin examination can be the key to an otherwise under-recognized cause of stroke

Keywords: Sneddon Syndrome; Livedo Racemosa; Ischemic Stroke; Thrombotic Vasculopathy; Cerebrovascular Disease

1 INTRODUCTION

Sneddon syndrome (SS) is a rare non-inflammatory arteriopathy characterized by the combination of livedo racemosa and cerebrovascular events, resulting from the thrombotic occlusion of small- and medium-sized arteries [1,2]. It was first described by Ian Bruce Sneddon in 1965 as the association of livedo reticularis and cerebrovascular [1]. The

estimated incidence is low, around four cases per million per year, and the syndrome predominantly affects women between the third and fifth decades of life [2,3].

The cutaneous hallmark is livedo racemosa, which differs from the more common livedo reticularis by its broken, irregular, and asymmetrical branching pattern, its wider distribution typically the trunk, buttocks, and proximal limbs, and its persistence on rewarming [2,4]. Whereas livedo reticularis reflects transient vasoconstriction, livedo racemosa reflects persistent impairment of cutaneous blood flow due to arterial occlusion [4]. Neurologically, SS manifests as recurrent transient ischemic attacks and ischemic strokes, and, over time, progressive cognitive decline [2,3].

Approximately half of patients have antiphospholipid antibodies, which allows a clinically useful distinction between antiphospholipid-antibody positive and negative forms of the syndrome [5]. The diagnosis is essentially clinical, supported by the characteristic skin findings, brain imaging, skin biopsy, and the exclusion of alternative causes of livedo and stroke [2,6]. Because the classic patient is young and female, SS is easily overlooked in elderly patients presenting with stroke, in whom cerebrovascular disease is more readily attributed to conventional vascular risk factors. We report a case of SS revealed by an ischemic stroke associated with livedo racemosa in a 69-year-old patient, to underline the value of a systematic skin examination in the stroke unit.

2 CASE PRESENTATION

A 69-year-old man was admitted to the neurology department for an acute ischemic stroke, presenting with the sudden onset of right-sided hemiplegia and aphasia. His medical history was notable for hypertension and type 2 diabetes mellitus, but he had no known history of prior stroke or transient ischemic attack, and no reported personal or family history of venous thromboembolism.

On admission, the patient was afebrile and hemodynamically stable, and neurological examination confirmed the right-sided hemiplegia and expressive aphasia. Dermatological examination revealed a widespread, persistent, net-like violaceous eruption with an irregular, broken, branching pattern, consistent with livedo racemosa, involving the trunk and lower limbs (Figure 1). The lesions were non-infiltrated and painless, without ulceration or nodules, and persisted on rewarming. The eruption was noted on admission and was of undetermined duration, as no reliable history of its onset could be obtained.

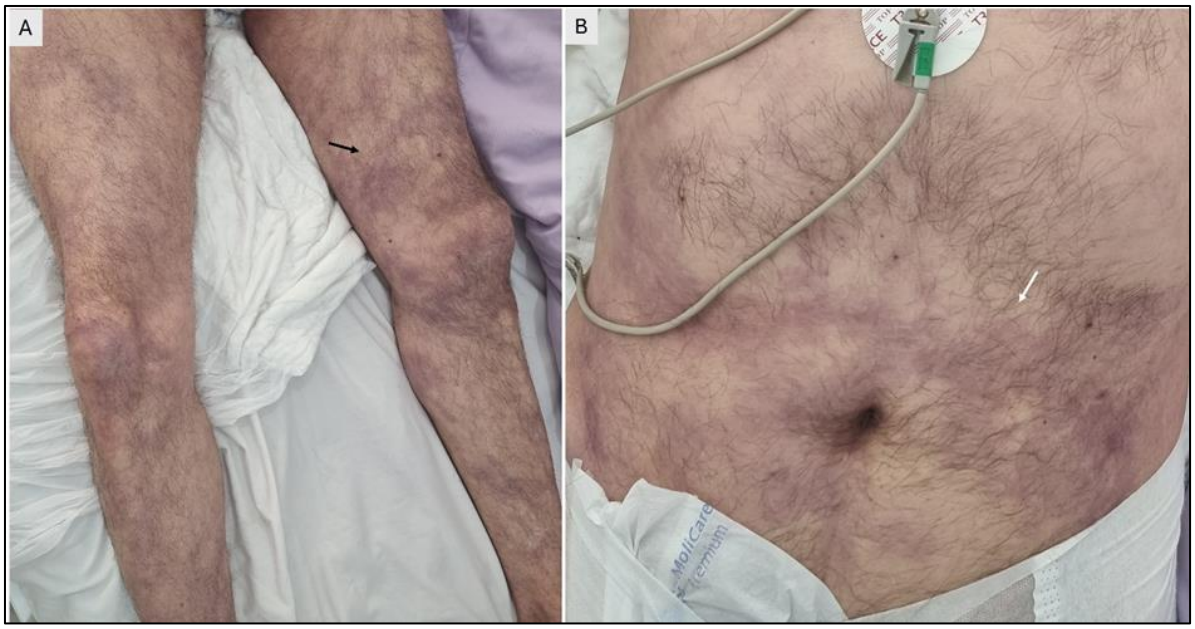

Figure 1: Clinical image showing widespread livedo racemosa with an irregular, broken, branching pattern, involving the lower limbs (black arrow, A) and trunk (white arrow, B).

Non-contrast brain computed tomography (CT) demonstrated an area of hypodensity in the left frontal cortico-subcortical region, within the territory of the left middle cerebral artery, consistent with a subacute ischemic infarction (Figure 2).

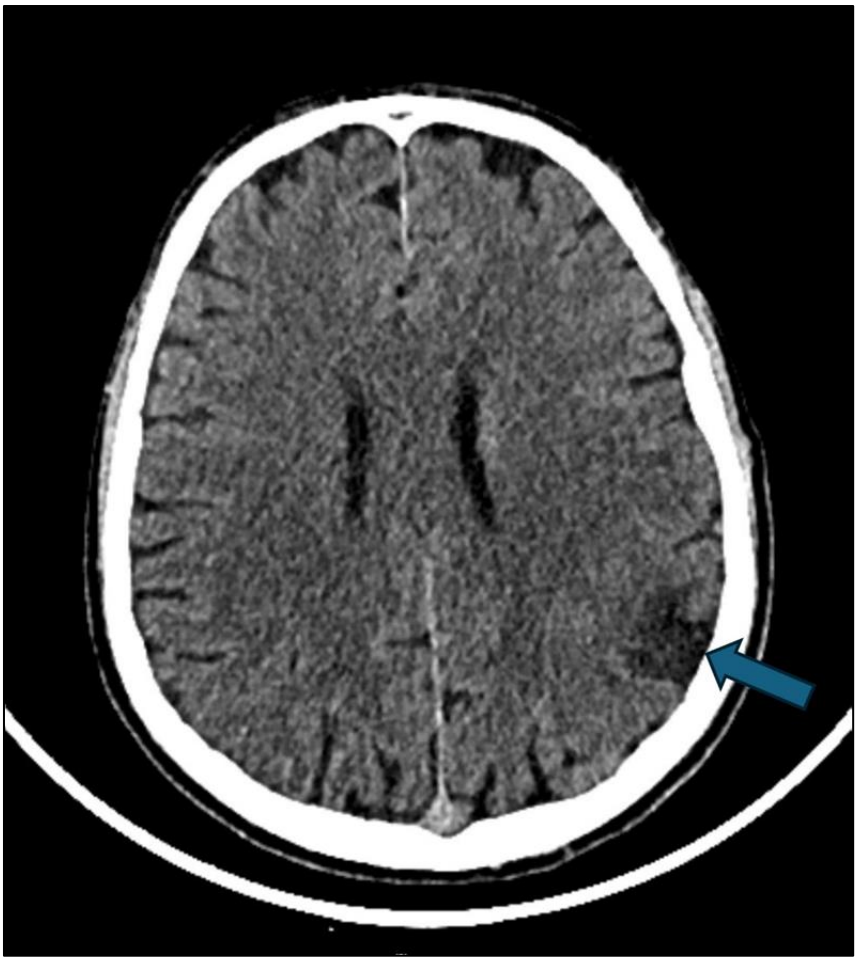

Figure 2: Non-contrast brain CT showing an area of hypodensity in the left frontal cortico-subcortical region (bleu arrow), within the territory of the left middle cerebral artery, consistent with a subacute ischemic infarction.

Initial laboratory investigations were within normal limits, with a hemoglobin level of 13.1 g/dL (13.0-17.0 g/dL), a white blood cell count of 8,100/mm^3 (4,000-10,000/mm^3), a platelet count of 260,000/mm^3 (150,000-400,000/mm^3), a C-reactive protein of 3.1 mg/L (<5 mg/L), and an erythrocyte sedimentation rate of 15 mm/h (<20 mm/h). The absence of an inflammatory syndrome was consistent with the non-inflammatory nature of the underlying vasculopathy. Cardiac work-up, including electrocardiography, transthoracic echocardiography, and Holter monitoring, was normal and did not reveal a cardioembolic source.

The immunological and thrombophilia work-up showed a negative antiphospholipid antibody profile, with a negative lupus anticoagulant, negative anticardiolipin antibodies (IgG and IgM), and negative anti-β2-glycoprotein I antibodies (IgG and IgM); antinuclear antibodies (ANA) were also negative. A skin biopsy was not performed, as the cutaneous eruption was clinically typical of livedo racemosa.

Based on the association of generalized livedo racemosa with a radiologically confirmed ischemic stroke, and after exclusion of alternative causes of livedo and stroke, a diagnosis of antiphospholipid-antibody-negative Sneddon syndrome was made. The patient was started on antiplatelet therapy with aspirin, together with optimization of vascular risk factors, including antihypertensive treatment, glycemic control, and lipid-lowering therapy with a statin. During the hospital stay, however, the patient's clinical condition deteriorated, with the onset of cardiogenic shock, to which he ultimately succumbed despite supportive management.

3 DISCUSSION

Sneddon syndrome is a rare thrombotic vasculopathy in which the occlusion of small- and medium-sized arteries produces the characteristic combination of livedo racemosa and cerebrovascular disease [1,2]. The pathophysiology remains incompletely understood; proposed mechanisms include a primary non-inflammatory endotheliopathy with progressive intimal proliferation and thrombotic occlusion, autoimmune mechanisms, and acquired or inherited thrombophilia, with a well-recognized association with antiphospholipid antibodies in roughly half of cases [2,5,6]. Rare

familial forms linked to NOTCH3 or CECR1/ADA2 mutations have also been described, supporting the heterogeneity of the syndrome [2].

The originality of our observation lies in the patient's age and sex. SS classically affects women between 30 and 50 years of age [2,3], and reports in men approaching 70 years are exceptional. In elderly patients with hypertension and diabetes, as in our patient, an ischemic stroke is readily attributed to atherosclerosis, small-vessel disease, or a cardioembolic source, and the cutaneous examination is often neglected. In this context, the recognition of a widespread livedo racemosa was the finding that redirected the diagnostic reasoning toward a systemic thrombotic vasculopathy. We acknowledge that the coexistence of conventional vascular risk factors makes the attribution of the stroke to SS more difficult, and that the two mechanisms may coexist; however, the presence of a typical, persistent, generalized livedo racemosa in a patient with an ischemic stroke fulfils the core clinical definition of the syndrome and warrants its recognition and specific follow-up [2,6]. This case therefore illustrates that a simple skin examination at the bedside can reveal an under-recognized cause of stroke even when common vascular risk factors are present.

Distinguishing livedo racemosa from the benign livedo reticularis is central to the diagnosis. Livedo reticularis produces a fine, regular, closed, symmetrical network that fades on rewarming and reflects transient vasoconstriction, whereas livedo racemosa is coarser, irregular, broken, asymmetrical, more widely distributed, and persists on rewarming because it results from fixed arterial occlusion [2,4]. This clinical distinction is more than semantic given that livedo racemosa is a marker of an underlying occlusive arteriopathy and, in the setting of arterial thrombosis, is strongly associated with antiphospholipid antibody syndrome [4].

The relationship between SS and the antiphospholipid syndrome (APS) deserves emphasis. In our patient, the antiphospholipid antibody work-up was negative. Importantly, a negative antibody profile does not exclude the diagnosis. Antiphospholipid-antibody-negative SS is well documented and defines a distinct subgroup with a clinical picture comparable to the antibody-positive form [5]. The 2023 ACR/EULAR APS classification criteria now formally incorporate non-thrombotic clinical features, including livedo racemosa, and require persistent antiphospholipid antibody positivity confirmed within a defined time window; our patient did not meet these criteria, which is consistent with an antibody-negative Sneddon syndrome rather than APS [7]. As a single negative determination does not fully rule out transiently positive or low-titer antibodies, repeat testing after an interval may be considered [7].

The diagnostic approach combines characteristic skin findings, brain imaging, skin biopsy, and the exclusion of differential diagnoses [2,6]. On MRI, ischemic lesions in SS tend to be small, multifocal, and located in the subcortical and periventricular white matter, and cerebral angiography is abnormal in a substantial proportion of patients [6]. Skin biopsy, ideally taken from the central pale area of the livedo network and including the deep dermis, may show the characteristic non-inflammatory occlusion of dermal arterioles, although its sensitivity is limited and multiple biopsies are sometimes required [6]. Differential diagnoses to exclude include systemic lupus erythematosus, polyarteritis nodosa, cryoglobulinemia, cholesterol embolism, cold agglutinin disease, and central nervous system vasculitis [2,6].

There is no standardized treatment for SS, and management is guided by the antiphospholipid antibody status and the individual thrombotic risk [2]. Antiplatelet agents are frequently used in antibody-negative patients, whereas long-term anticoagulation is generally favored in antiphospholipid-antibody–positive patients or after recurrent ischemic events; strict control of cardiovascular risk factors is recommended in all cases [2,5]. In our antibody-negative, elderly patient, antiplatelet therapy combined with aggressive control of hypertension, diabetes, and dyslipidemia was chosen, a strategy that also avoided the higher hemorrhagic risk associated with long-term anticoagulation at this age. Despite this management, the patient died from cardiogenic shock. This fatal outcome is a reminder that SS is a systemic thrombotic arteriopathy that is not confined to the skin and brain but may also involve the coronary and cardiac vasculature, and that cardiovascular events contribute to its morbidity and mortality, particularly in older patients with additional vascular risk factors [2,5].

This report has several limitations. It describes a single case, and the antiphospholipid antibody profile was based on a single determination that could not be repeated after the recommended interval of at least 12 weeks because the patient died during the hospitalization [7]. A skin biopsy, which can provide histological support by showing non-inflammatory occlusion of dermal arterioles, was not performed; the diagnosis rested on the clinically typical appearance of the livedo racemosa combined with the radiologically confirmed stroke and the exclusion of alternative causes, an approach consistent with the essentially clinical nature of the diagnosis [2,6]. Negative antinuclear antibodies argued against an associated connective-tissue disease such as systemic lupus erythematosus. Finally, the coexistence of hypertension and diabetes represents an alternative contributor to the cerebrovascular event. These limitations notwithstanding, the case adds to the limited literature on SS in elderly men and reinforces the message that the skin is a valuable diagnostic window in the stroke unit.

4 CONCLUSION

Sneddon syndrome is a rare thrombotic vasculopathy that should be considered in any patient presenting with an ischemic stroke associated with livedo racemosa, including elderly patients in whom the diagnosis is easily missed. This case illustrates how a simple, systematic skin examination in the stroke unit can reorient the etiological work-up and reveal an underlying systemic arteriopathy. Recognition of livedo racemosa, appropriate antiphospholipid antibody testing with confirmation over time, and exclusion of alternative causes are the cornerstones of the diagnosis. Early identification is important because it guides antithrombotic management and secondary prevention, with the aim of reducing the risk of recurrent cerebrovascular events.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

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Disclosure of conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

Statement of informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. Institutional ethical approval was not required for a single case report according to local regulations.

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