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# Sneddon Syndrome, About a Case Report and Literature Review

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**Abstract** **Case Report**

Sneddon's syndrome is a rare condition characterized by livedo and multiple ischemic strokes with no apparent cause. This report presents a case of a 34-year-old woman with a 16-year history of livedo who developed right hemibody heaviness, a decrease in visual acuity, and palpitations. The MRI showed bilateral parieto-occipital cortico-subcortical atrophy associated with abnormal signals in the deep white matter, parieto-occipital gliosis foci, and small-caliber artery vasculitis. The patient was eventually diagnosed with Sneddon's syndrome based on her clinical presentation, MRI results, and symptom evolution. The underlying mechanisms of the syndrome remain unclear, but it is thought to involve occlusion of small- and medium-sized arterioles. The neurological involvement of Sneddon's syndrome is highly diverse and dominated by cerebral ischemic events, leading over time to intellectual deterioration or even dementia. This report highlights the importance of considering Sneddon's syndrome as a differential diagnosis in cases of livedo and unexplained cerebral ischemia.

**Keywords:** Sneddon's syndrome, livedo, ischemic strokes.



## INTRODUCTION

Sneddon's syndrome is a rare disease characterized by the presence of extensive livedo and multiple ischemic strokes without an apparent cause. The first reported case was by Champion and Rook in 1960 [1]. Five years later, this entity was named after Dr. Ian Bruce Sneddon, a British dermatologist who considered this pathological association as a distinct disease by reporting six cases [2].

Sneddon's syndrome is an extremely rare medical condition, affecting approximately 4 people per million, and primarily affecting women aged 30 to 40 years [3]. The exact mechanisms behind this disease are not yet fully understood, but some hypotheses have been explored. The clinical presentation of Sneddon's syndrome is actually quite homogeneous, with livedo preceding cerebrovascular involvement by several years, and dementia, if it occurs, by several decades. However, it is often misdiagnosed as central nervous system vasculitis [4].

Medical imaging, mainly MRI, plays an important role in the positive diagnosis of this entity, if the radiological characteristics are taken into account, thus avoiding the administration of unnecessary treatments that could be potentially dangerous [5].

We report here the case of a 34-year-old woman in whom Sneddon's syndrome was diagnosed 16 years after the onset of livedo.

## OBSERVATION

This is a 34-year-old woman with a medical history of intrauterine fetal death with miscarriage and repeated transient ischemic attacks that were not explored. The patient was hospitalized in the dermatology department for livedo evolving for 16 years, with the recent onset and worsening of right hemibody heaviness, associated with a decrease in visual acuity and palpitations.

The clinical examination showed a conscious patient who was stable on the cardiovascular and respiratory level. The skin was smooth with non-infiltrated and irregular livedo racemosa (Figure 1), accentuated in the standing position without detectable necrotic or purpuric lesions. Visual acuity was estimated to be 4/10 in the right eye and 10/10 in the left eye without detectable abnormalities on fundoscopy.

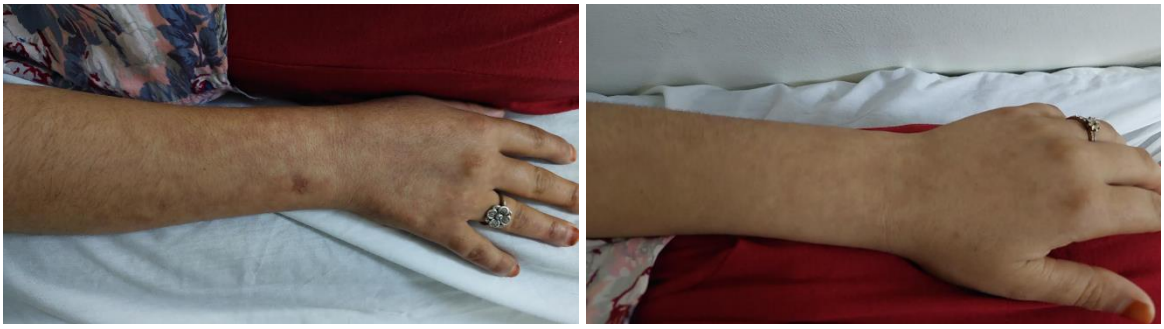

**Figure 1: Livedo Raemosa in both hands of the patient**

The neurological examination revealed right hemibody heaviness with decreased muscle strength and hypoesthesia. The laboratory workup showed a normal complete blood count with an estimated platelet count of 270,000, negative CRP, normal liver function and coagulation tests, and negative serologies (for syphilis, HIV, and hepatitis). Cardiac exploration by ECG and echocardiography showed no abnormalities

An MRI was requested to explore the probable etiology of the right hemispheric heaviness and decrease in visual acuity. The MRI was performed with and without contrast injection, including angio-MRI sequences. The exploration revealed bilateral parieto-occipital cortico-subcortical atrophy associated with abnormal signals in the deep white matter above and below the tentorium, and bilateral parieto-occipital gliosis foci as a sequelae of previous infarctions (Figure 2 & 3).

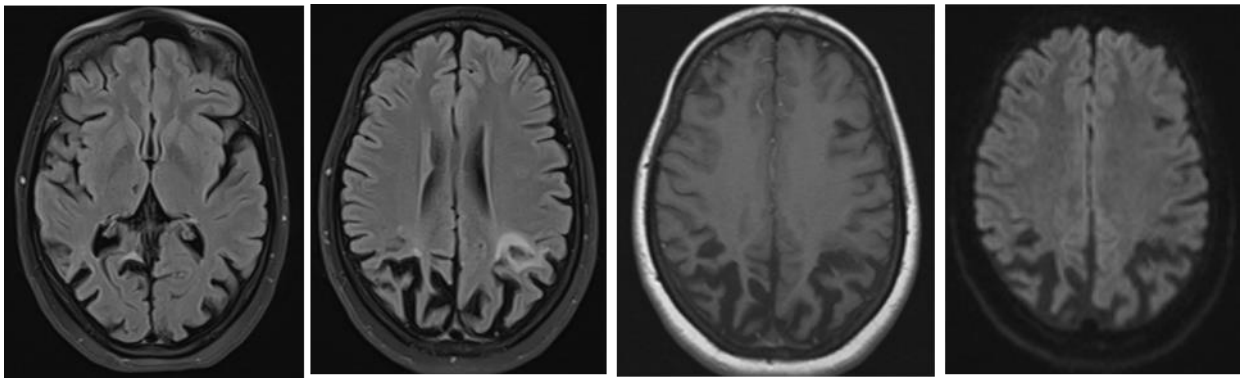

**Figure 2: Axial Flair and T1 brain MRI sequence, showing bilateral parieto-occipital cortico-subcortical atrophy with white matter signal abnormalities suggestive of sequela of ischemic changes**

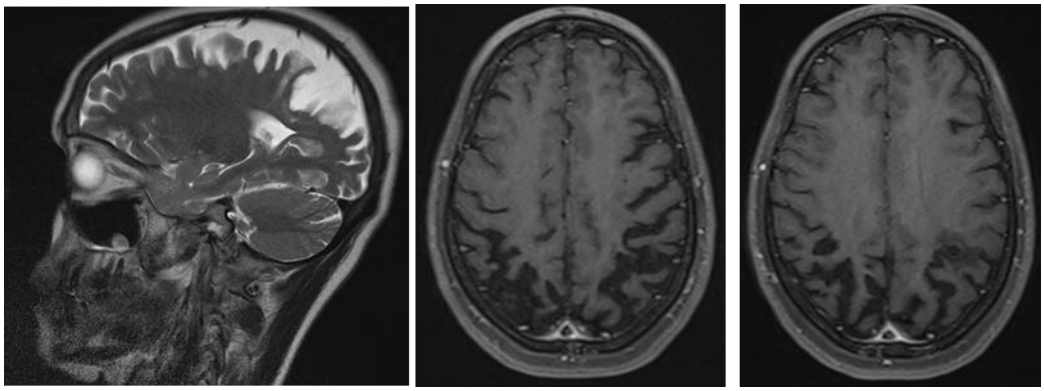

**Figure 3: Brain MRI T2 sagittal sequence and T1 with gadolinium injection showing white matter signal abnormalities not enhanced after injection**

The major vascular axes were normal on the angio-MRI sequences. The overall lesions observed on the MRI were suggestive of a vasculitis affecting mainly small-caliber arteries (Figure 4).

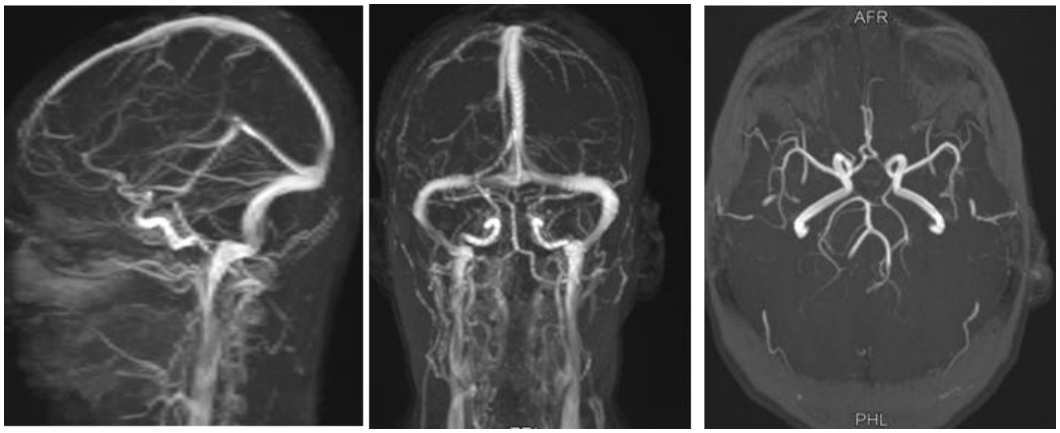


**Figure 4: Cerebral MR angiography without gadolinium injection: Non-visualization of small and medium-sized arteries supplying the posterior brain**

Given this situation, neuro-lupus was considered due to the history of fetal death and miscarriage, but negative serologies and immunological testing ruled out this diagnosis. Other vasculitides such as Wegener's granulomatosis were also considered, but the clinical and biological presentation was not suggestive. Due to the presence of livedo for 16 years, Sneddon's syndrome was considered despite its rarity, but the clinical presentation, MRI results, and evolution of symptoms led to a diagnosis with more certainty than the previously described pathological entities.

## DISCUSSION

Sneddon syndrome (SS) is a rare non-inflammatory thrombotic vasculopathy characterized by the combination of cerebrovascular disease with livedo racemosa [6]. Its incidence is estimated at 4 cases per million inhabitants per year, and it is sporadic, although rare familial cases have been reported with autosomal dominant or recessive transmission [7].

The exact mechanism of Sneddon's disease is not fully understood. However, it is generally accepted that the occlusion of small- and medium-sized arterioles is involved, which can cause skin lesions and affect the central nervous system, as well as the renal arterioles to a lesser extent. Some have suggested a vascular origin with obliterative endarteritis, while others have proposed an embolic mechanism. The association with antiphospholipid antibodies has led to suspicion of an immunological thrombotic process, but this does not explain all cases of the disease [8].

The Sneddon syndrome can be either primary or considered a common clinical manifestation of different pathological entities. Frances et al. [9] proposed that the Sneddon syndrome can be classified into three categories depending on its association with antiphospholipid syndrome or systemic lupus erythematosus. Non-infiltrating livedo racemosa is the main clinical manifestation of this disease, mainly located on the limbs (100%), but also on the trunk (84-89%), buttocks (68-74%), extremities (53-59%), and face (15-16%) [6]. Generally, this manifestation precedes neurological symptoms by several years, but in some cases, it can appear simultaneously or later.

The neurological involvement of Sneddon syndrome is highly diverse and dominated by cerebral ischemic events (CIEs), whose clinical expression depends on the location of the ischemic lesion. Ataxia and/or language disorders can be observed. The repetition of these CIEs can lead over time to intellectual deterioration or even dementia, sometimes as an inaugural symptom if the infarcts are silent [10]. Seizures reflecting cortical involvement with hemorrhagic strokes have been described but rarely observed [11]. In addition to the two cardinal symptoms, skin and neurological, arterial hypertension, valvulopathies, ocular involvement, and renal impairment have been reported, completing the clinical picture of Sneddon syndrome.

The biological assessment in Sneddon syndrome is not specific and may show an inflammatory biological syndrome with a high erythrocyte sedimentation rate (ESR). The positivity of anti-phospholipid antibodies is highly variable among authors and ranges from 0 to 85% [6]. Our case presented typical clinical manifestations of Sneddon syndrome associated with other non-specific symptoms such as palpitations and decreased visual acuity. The latter indicates the chronicity of the condition as well as the insidious evolution of the disease dating back more than 15 years.

Cerebral MRI is considered an essential examination in the diagnosis of Sneddon syndrome. The lesions have similar characteristics with a typical appearance that includes multiple medium to small cortico-subcortical infarcts and a non-territorial cerebellar infarct on a background of diffuse and mild focal cerebral atrophy. In a study of radiological characteristics in 12 patients with Sneddon syndrome, cerebral MRI showed that almost all cortico-subcortical infarcts were located in the supratentorial regions, and

all patients had at least one non-territorial cerebellar infarct. Cerebral microbleeds are very rare and are often associated with hypertensive phenomena [5].

The diffusion sequence never shows recent embolic patterns in patients with Sneddon syndrome. Additionally, involvement of the white matter is mild, and the basal arteries and perforators are not primarily involved in the disease. Finally, diffuse cerebral atrophy is mild, but there may be significant focal tissue loss as a sequelae of covered infarcts leading to disabling neurological dysfunction and epilepsy [5, 12].

The presence of these almost typical findings on MRI in a young woman without significant risk factors should always lead us to consider the diagnosis of Sneddon syndrome. If livedo is detected, the diagnosis will be established, although there are cases in the relevant literature where livedo is diagnosed after a stroke [13].

Our patient presented with livedo preceding neurological involvement with MRI characteristics that are almost typical of those reported in the literature.

## CONCLUSION

Sneddon's syndrome is a very rare clinical syndrome that likely results from a number of acquired or congenital hemostatic abnormalities that preferentially affect the cerebral and cutaneous vascular beds. The low number of reported cases may partly reflect a lack of knowledge of the syndrome rather than its true incidence. While the radiological characteristics are generally identified and studied, it appears that further research into the pathogenesis of Sneddon's syndrome is necessary to identify its etiological subgroups as well as their therapeutic modalities.

## REFERENCE


1. Champion, R. H. (1960). Livedo reticularis. In *Proc R Soc* (Vol. 53, pp. 961-962).
2. Sneddon, I. B. (1965). Cerebro-vascular lesions and livedo reticularis. *British Journal of Dermatology*, *77*(4), 180-185.
3. Bottin, L., Francès, C., de Zuttere, D., Boëlle, P. Y., Muresan, I. P., & Alamowitch, S. (2015). Strokes in S neddon syndrome without antiphospholipid antibodies. *Annals of neurology*, *77*(5), 817-829.
4. Samanta, D., Cobb, S., & Arya, K. (2019). Sneddon syndrome: a comprehensive overview. *Journal of Stroke and Cerebrovascular Diseases*, *28*(8), 2098-2108.
5. Yilmaz, E., Arsava, E. M., Gocmen, R., Oguz, K. K., Arat, A., & Topcuoglu, M. A. (2021). Characteristic imaging features of neurovascular involvement in primary Sneddon's syndrome: an analysis of 12 cases. *Neurological Sciences*, *42*, 2363-2369.
6. Wu, S., Xu, Z., & Liang, H. (2014). Sneddon's syndrome: a comprehensive review of the literature. *Orphanet journal of rare diseases*, *9*(1), 1-7.
7. Lossos, A., Ben-Hur, T., Ben-Nariah, Z., Enk, C., Gomori, M., & Soffer, D. (1995). Familial Sneddon's syndrome. *Journal of neurology*, *242*, 164-168.
8. El Benaye, J., Hsaini, Y., Zoobo, T., Ghfir, M., Bourazza, A., & Sedrati, O. (2013). Le syndrome de Sneddon. *La Presse Médicale*, *42*(2), 138-144.
9. Francès, C., Papo, T., Wechsler, B., Laporte, J. L., Biousse, V., & Piette, J. C. (1999). Sneddon syndrome with or without antiphospholipid antibodies: a comparative study in 46 patients. *Medicine*, *78*(4), 209-219.
10. Lavernhe, S., Richard, A., Varvat, J., Garnier, P., Dirson, S., & Thomas-Antérion, C. (2013). Le syndrome de Sneddon: une cause rare de démence vasculaire. *Revue Neurologique*, *169*(2), 169-172.
11. Bayrakli, F., Erkek, E., Kurtuncu, M., & Ozgen, S. (2010). Intraventricular hemorrhage as an unusual presenting form of Sneddon syndrome. *World neurosurgery*, *73*(4), 411-413.
12. Starmans, N. L. P., van Dijk, M. R., Kappelle, L. J., & Frijns, C. J. M. (2021). Sneddon syndrome: a comprehensive clinical review of 53 patients. *Journal of Neurology*, *268*, 2450-2457.
13. Cavestro, C., Richetta, L., Pedemonte, E., & Asteggiano, G. (2009). Sneddon's syndrome presenting with severe disabling bilateral headache. *The Journal of headache and pain*, *10*(3), 211-213.