Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Sneddon Syndrome: A Case Report

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DOI: 10.36347/sjmcr.2024.v12i03.023 | **Received:** 02.02.2023 | **Accepted:** 14.03.2024 | **Published:** 27.03.2024

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Abstract **Case Report**

Sneddon syndrome is a rare autoimmune disorder characterized by a non-inflammatory arteriopathy that affects small and medium-sized arteries. The condition is challenging to diagnose due to its nonspecific clinical manifestations. This case report presents a 34-year-old male patient with sudden-onset visual acuity loss in the left eye and progressive weakness of the left hemibody, associated with spastic tetraparesis and gait disturbance. The clinical presentation and imaging findings were consistent with Sneddon syndrome. MRI played a crucial role in the diagnosis, revealing asymmetrical cortical and subcortical signal abnormalities, periventricular white matter nodules, and lacunar infarcts. These findings are consistent with the known radiologic features of Sneddon syndrome. The case emphasizes the importance of imaging techniques, particularly MRI, in the diagnosis and management of Sneddon syndrome. It also highlights the challenges in diagnosing Sneddon Syndrome and the importance of considering this condition in young patients presenting with neurological symptoms and skin lesions.

Keywords: Sneddon syndrome; arteriopathy; livedo racemosa.

INTRODUCTION

Sneddon syndrome is a rare autoimmune disorder, which is considered a non-inflammatory arteriopathy that affects small and medium-sized arteries, characterized by the presence of livedo racemosa, skin condition characterized by a net-like or lace-like pattern of red or blue discoloration of the skin, and cerebral thrombosis which can lead to stroke or transient ischemic attacks (TIAs) [2].

The condition was first described by Sneddon in 1965 and is believed to affect approximately 4 per million individuals.

The diagnosis of Sneddon syndrome is often challenging due to the nonspecific clinical manifestations. However, imaging techniques, including magnetic resonance imaging (MRI), have proved to be valuable tools in the diagnosis of Sneddon syndrome [2]. MRI can reveal characteristic findings such as hyperintense lesions in the white matter, lacunar infarcts, and atrophic changes in the optic nerve. In this context, we present a case of a 34-year-old man with a sudden onset of visual acuity loss in the left eye and progressive weakness of the left hemibody, associated with spastic tetraparesis and gait disturbance. The clinical presentation and imaging findings were consistent with Sneddon syndrome.

Through this case report, we emphasize the importance of MRI in the diagnosis of Sneddon syndrome and its potential role in guiding the management of this rare disease.

OBSERVATION

The case report is about a 34-year-old male patient, presented with a sudden onset of decreased visual acuity in his left eye that had been ongoing for the past five years, without any associated signs. One year later, he developed progressive functional impairment of the left side of his body and gait disturbance, in the absence of fever or other systemic symptoms. On clinical examination, he had spastic tetraparesis and bilateral visual acuity loss, along with livedo racemosa skin lesions

Laboratory investigations, including a workup for systemic diseases, vasculitis, and viral infections such as hepatitis, HIV, and syphilis, were unremarkable. A lumbar puncture was performed, which revealed no abnormalities. An electrocardiogram and echocardiography also showed no abnormalities.

An MRI of the brain was performed, which revealed asymmetrical cortical and subcortical signal abnormalities in a gyriform pattern, involving bilateral fronto-parietal and occipital regions, seen as T2 and FLAIR hypersignals, without diffusion restriction or contrast enhancement. Additionally, nodular T2 and FLAIR hypersignals were seen in the periventricular white matter. Multiple lacunar lesions were also noted in the supra- and infratentorial regions, seen as T2 hypersignals that disappeared on FLAIR (Figure 1: A B C D E F G H). These findings are consistent with the diagnosis of Sneddon Syndrome. The MRI also revealed optic nerve atrophy on the left side, particularly involving its intraorbital and proximal intracanalicular segments.

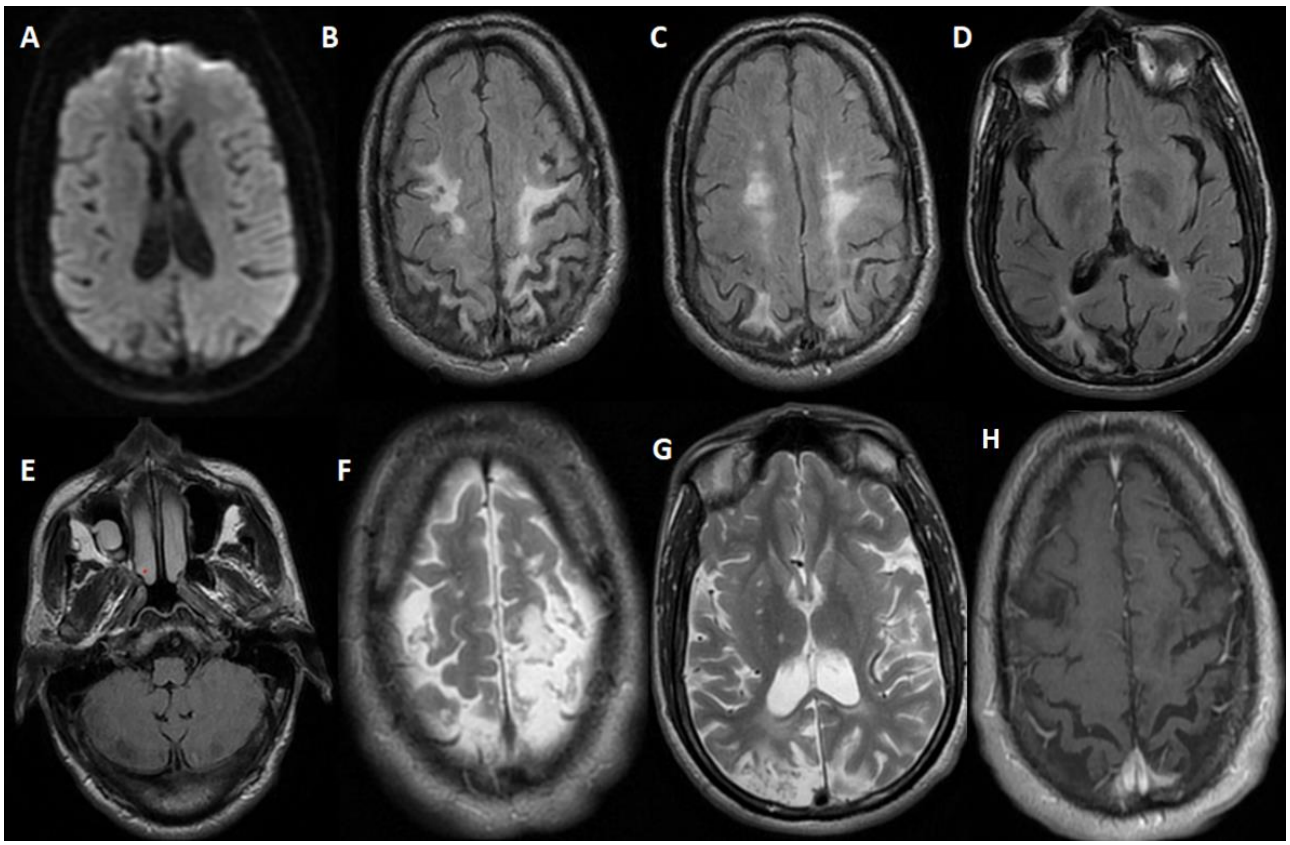

Figure 1: A) DWI showing no abnormalities; B,C,D,E) axial FLAIR WI showing asymmetrical cortical and subcortical signal abnormalities in a gyriform pattern, involving bilateral fronto-parietal and occipital regions; F,G axial T2 WI showing multiple lacunar lesions; H axial T1 SE no enhancement lesions

DISCUSSION

This case highlights the challenges in diagnosing Sneddon Syndrome and the importance of considering this condition in young patients presenting with neurological symptoms and skin lesions.

Imaging studies, particularly MRI, play a crucial role in the diagnosis of Sneddon syndrome.In our case, MRI revealed cortical and subcortical signal abnormalities, periventricular white matter nodules, and lacunar infarcts. These findings are consistent with the known radiologic features of this disorder. The cortical and subcortical signal abnormalities, often in a gyriform pattern, are thought to represent ischemic changes related to thrombotic events in the cerebral vasculature [1]. The periventricular white matter nodules, also known as Lhermitte-Duclos lesions [2], are thought to be a result of infarction or edema of the periventricular white matter. The lacunar infarcts, which are small infarcts in the deep brain structures, are thought to result from small vessel disease.

Our findings are consistent with those reported by other authors in the literature. For example, Cavestro *et al*., [3] reported similar radiologic findings in a case of Sneddon syndrome, including cortical and subcortical signal abnormalities, periventricular white matter nodules, and lacunar infarcts. Similarly, Yilmaz *et al.*, [4] reported similar MRI findings in a case of Sneddon syndrome, including cortical and subcortical signal abnormalities and periventricular white matter lesions.

In a case series by Cleaver *et al.*, [6] MRI was used to identify radiologic features of Sneddon syndrome in four patients. These features included cortical and subcortical signal abnormalities, periventricular white matter nodules, and lacunar infarcts. In addition, Aladdin [7] reported similar MRI findings in a case of Sneddon

syndrome, including multiple infarcts and periventricular white matter abnormalities.

Magnetic Resonance Imaging (MRI) plays a pivotal role in the diagnosis of Sneddon's syndrome, a rare and often underdiagnosed vascular disorder. The distinctive cerebral microangiopathy seen on MRI, characterized by multiple ischemic lesions affecting both grey and white matter, periventricular white matter nodules, and lacunar infarcts, serves as a hallmark feature [5]. Additionally, MRI can aid in differentiating Sneddon's syndrome from various potential mimickers, such as multiple sclerosis, systemic lupus erythematosus, and other autoimmune or vasculitic conditions [4]. Its ability to provide non-invasive, high-resolution images of the brain and vascular system makes MRI an invaluable tool for not only confirming the diagnosis but also for ruling out alternative pathologies, ultimately guiding clinicians toward appropriate management strategies for this challenging condition.

Additionally, laboratory investigations may include coagulation studies, autoimmune markers, and genetic testing to rule out other systemic and thrombotic disorders. Confirmation of Sneddon's syndrome often relies on the presence of characteristic histopathological findings in skin biopsies, demonstrating arteriolar occlusion and endothelial proliferation. Despite advancements in diagnostic modalities, early recognition and multidisciplinary collaboration remain crucial for timely management and improved outcomes in patients with Sneddon's syndrome [3].

The management of Sneddon's syndrome represents a complex and evolving challenge in the realm of vascular medicine. Currently, therapeutic interventions focus on two primary aspects: the prevention of new ischemic events and the management of associated comorbidities. Antiplatelet agents, particularly aspirin, have shown efficacy in reducing the risk of recurrent strokes or ischemic events in some patients. In more severe cases or when antiplatelet therapy is inadequate, anticoagulation with agents such as warfarin or direct oral anticoagulants may be considered. Additionally, addressing underlying risk factors such as hypertension and hyperlipidemia is paramount [4]. Despite these efforts, it is essential to acknowledge the variable and unpredictable course of Sneddon's syndrome. The long-term prognosis remains guarded, with some individuals experiencing a relatively stable clinical course, while others may face progressive vascular involvement, leading to significant disability. A multidisciplinary approach, involving neurologists, dermatologists, radiologists and vascular specialists, is crucial for tailoring treatment strategies to individual patient needs and monitoring disease progression over time [7].

CONCLUSION

Sneddon Syndrome is a rare condition that can present with a wide range of neurological and dermatological manifestations [1]. This case report emphasizes the importance of considering Sneddon Syndrome in the differential diagnosis of young patients with neurological symptoms and skin lesions, and highlights the role of MRI in supporting the diagnosis [4].The imaging findings on MRI, including cortical and subcortical signal abnormalities in a gyriform pattern and optic nerve atrophy, are characteristic of Sneddon Syndrome and can aid in the diagnosis [6]. Early recognition and treatment with anticoagulation therapy can improve outcomes and prevent further cerebrovascular events [3]. Clinicians should remain vigilant for this rare but potentially serious condition in patients with compatible clinical features and imaging findings [7].

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