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Scleroderma with Neurological System Involvement: A Case Report

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CASE REPORT

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Ethics Approval
After the patient is informed about the study, oral and written consent is obtained and confidentiality of private information about the patient is ensured. It is stated that it will be protected.

Conflict of Interest
There is no conflict of interest.

Author Contribution
Idea, concept and design: MK
Data collection and analysis: MK
Drafting of the manuscript: MK
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Data Availability
Data supporting the findings of this study are available upon reasonable request can be obtained from the corresponding author.
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A B S T R A C T

Scleroderma is a rare autoimmune disease characterized by inflammation, vascular damage and fibrosis. The importance and frequency of neurological involvement in scleroderma is a matter of significant debate in the literature. In the past years, it has been reported that central nervous system involvement is unusual in systemic scleroderma. Determinants of neurological damage have been shown to be renal crises, cardiovascular and pulmonary involvement, and treatment. In recent years, studies based on magnetic resonance imaging describe changes often associated with asymptomatic neurological involvement, regardless of disease severity, complications, or duration. In this case report, a case of neurological involvement with ataxia, which is rare in scleroderma, was examined.

Keywords: scleroderma, neurological involvement, ataxia, asymptomatic neurological involvement

Scleroderma is a connective tissue disease that causes fibrosis in the skin, visceral organs and vascular system. Scleroderma occurs in two forms: localized and systemic. In localized form (morphea), skin and subcutaneous tissue involvement, sometimes deeper tissues may also be involved. In the systemic form, there is fibrosis in the skin and internal organs, including blood vessels, gastrointestinal tract, lungs, heart and kidneys (Signh et al., 2019). Central and peripheral nervous system involvement poses many diagnostic difficulties. In previous approaches, it was assumed that changes in the nervous system in systemic sclerosis were rare or secondary to other organ changes such as vasculitis, renal involvement, hypoxemia, pulmonary. Recent studies suggest that nervous system (NS) changes in systemic scleroderma may be directly related to the disease. Recent studies conducted with magnetic resonance imaging have shown asymptomatic central nervous system involvement, regardless of the severity and complications of scleroderma. It is hypothesized that vascular lesions with symptoms identical to those in Raynaud's phenomenon may lead to nervous system involvement in the brain and peripheral small vessels that vascularize the peripheral nerves(Sobolewski et al., 2019) . The most common symptoms of neurological involvement in systemic scleroderma are headache and dizziness, convulsions, visual disturbances and aphasia. Less common symptoms include anxiety, depression, psycho-organic syndromes, cognitive and orientation disorders, and even loss of consciousness (Golden and Vernino, 2019).

Depending on the stage of the disease, transient ischemic attack (TIA) and/or other ischemic syndromes, as well as stroke, trigeminal neuroinflammation, peripheral polyneuropathy and cranial nerve inflammation may also be observed. In the early stages, attacks of ischemic stroke, TIA or cerebral hemorrhage are observed. In the long term, it causes memory and mental disorders as well as depressive attacks. As neuroradiological methods, magnetic resonance imaging (MRI), computed

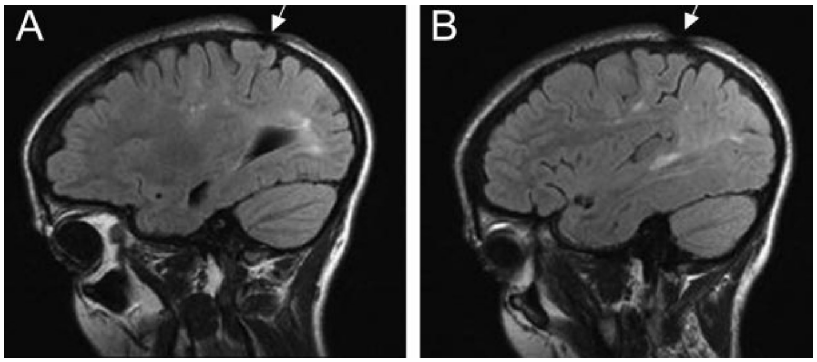

Figure 1. Multifocal high signal in periventricular and cortical-subcortical areas (Representative sagittal FLAIR images) Scleroderma is accompanied by focal atrophy of subcutaneous tissues.

tomography (CT), single photon emission computed tomography (SPECT) and positron emission tomography (PET) are used in the diagnosis of NS damage in systemic scleroderma. Peripheral nerve involvement is determined by nerve conduction studies. Demonstration of the nature of peripheral nerve involvement and differentiation of neurogenic and myogenic lesions, polyneuropathy and mononeuropathy are provided by Electromyography (EMG) (Desbois and Cacoub, 2016).

The diagnosis of scleroderma is determined clinically by the presence of Raynaud phenomenon, typical skin thickening and internal organ involvement. Laboratory examinations are also supportive. Serology testing for antibody profiles is useful to identify disease subtypes or to rule out conditions that mimic scleroderma. During the course of the disease, examinations are carried out to determine the extent and stage of involvement of internal organs (Lefaan et al., 2021).

Treatment of scleroderma is difficult due to the variability of disease symptoms and the cumulative effect of progressive fibrosis, obliterative vascular changes, and immune system activation and autoimmunity. Treatment of scleroderma is directed at categories that include medications, herbs, antioxidants, and biologics. The new delivery system-based approach generally stands out among other approaches consisting of vesicular systems, particulate systems, self-assembling systems, and eukaryotic and prokaryotic cell carriers. The goal of treatment is to prevent inflammation, vascular damage, excessive collagen production and fibrosis (Amaral et al., 2013).

In this case report, the patient's condition regarding neurological involvement, which is rare in patients diagnosed with Scleroderma, will be discussed.

CASE REPORT

A 37-year-old female patient applied to the emergency department with complaints of dizziness, loss of balance, and numbness and tingling in her fingers. Neurological examination revealed a glove-sock-like sensory deficit and a positive Romberg test. The sole of the foot reflexes were unresponsive, and deep tendon reflexes could not be obtained in any focus. Other neurological examination findings were normal. Carotid and vertebral Doppler ultrasonography was normal. Findings consistent with cerebellar atrophy were observed in brain tomography and magnetic resonance imaging (Figure 1). Diffuse sensorimotor peripheral neuropathy was detected on electromyography. Cerebellar ataxia was considered in the patient.

In her labarotory findings for etiology, hemogram, kidney and liver function tests, vitamin B12 and thyroid function tests were normal (Table 1). Tests for possible viral and bacterial agents were negative. No pathology was detected in the cerebrospinal fluid (CSF) examination. There was no growth in CSF culture (Table 2). The autoantibodies evaluated were negative except for ANA. Scl-70 was positive in the ENA profile.

Table 1. Laboratory Findings

Hb: 13,4 g/dl	**BUN:** 14	**Calsium:** 9,8 mg/dl	**Brucella:** Negative
Plt : 264,000	**Creatinine:** 0,96 mg/dl	**Albumin:** 4,5 g/dl	**Hepatit serologia:** Negative
WBC: 9700	**AST:** 11	**Magnezyum:** 1.9 mg/dl	**Fasting blood sugar:** 92 mg/dl
TSH: 2.6 uıU/ml	**ALT:** 29	**Sedimentation:** 24 mm/h	**Full urinalysis:** Normal
Ferritin: 98 ng/ml	**Folik asit:** 13.54 ng/ml	**CRP:** 0,44 mg/L	**Vit B12:** 361 pg/ml

Table 2. Cerebrospinal Fluid Examınatıon
Glucose: 67
Micrototal protein: 62.6 mg/dl (15-40)
LDH: 24 u/L (100-210)
Na: 150 (27-147)
CSF Cytology: benign CSF cytology containing a few PMNL-Lymphocytes

In her rheumatological evaluation, the patient stated that she had dry mouth and eyes, and it was observed that there was hardening of the skin on her face, arms and legs, facial lines disappeared and mouth opening decreased. The Reynould phenomenon was positive. Nail bed videcapillaroscopy was compatible with scleroderma.

Neurological involvement of scleroderma was considered in the patient. Steroids and sclophosphamide were started. It was observed that neuropathy and ataxia symptoms improved in the 6th month of treatment.

DISCUSSION

Neurological involvement is rare in scleroderma. Therefore, case reports are very important in terms of symptoms and treatment approaches of the disease. In a study examining the neurological symptoms of 42 patients with systemic sclerosis, it was determined that the rate of tonic-clonic convulsive epileptic attacks was 42%(Amaral et al., 2013). The diagnoses of the patients were mostly determined after these attacks. However, in this case, the dizziness is a distinct and obvious occurrence in the manipulation of the feet and the deep tendon reflex. Additionally, CSF is normal in the case. Oligoclonal IgG is frequently detected in past studies (Prajjval et al., 2024). Cerebellar ataxia has been reported rarely, and in this respect, our case report stands out by emphasizing a rare symptom in systemic scleroderma (Polunosika et al., 2022). It may also be beneficial for researchers and clinicians to consider systemic scleroderma in patients presenting with cerebellar ataxia.

CONCLUSION

Neurological involvement in scleroderma has begun to be detected with increasing frequency in recent years. Since it is mostly associated with disease activity, it stands out as a striking symptom in scleroderma, considering that possible permanent damage can be prevented by early detection and treatment. This study has limitations in generalizing it to the literature as it is the report of a single case. However, it is very important as it will remind you to consider the diagnosis of systemic scleroderma when encountering a patient with neurological symptoms such as dizziness, loss of balance, and cerebellar ataxia. For future studies, it is thought that the case report will contribute to the examination of neurological symptoms related to rare systemic scleroderma and the inclusion of case series in the literature.

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