NEUROLOGICAL INVOLVEMENT IN THE COURSE OF SCLEROMYXEDEMA: A CASE REPORT
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INTRODUCTION Scleromyxedema is a rare disease of unknown etiology primarily affecting the skin, characterized by generalized papular eruption, dermal fibroblast proliferation with mucin deposition, and a monoclonal gammopathy [1]. It usually affects middle-aged individuals and frequently presents extracutaneous complications with multiorgan involvement [2]. Neurological impairment is a rare but sometimes fatal complication of scleromyxedema that should be rapidly identified to prevent significant morbidity and mortality.

CASE REPORT A 63-year-old caucasian man had a two-year history of Scleromyxedema, based on the characteristic skin papular eruption (Figure 1), skin biopsy consistent with mucinosis and a monoclonal gammopathy. Immunosuppressive treatment with cyclosporine and methotrexate was started and carried on since the diagnosis, with benefit. He came to our attention because of sudden alterations in neurological function following a two days period of flu-like symptoms. According to his relatives he was sleepy, confused and could not speak clear words. After few hours since admission he developed left hemiparesis. The brain computed tomography and cerebrospinal fluid (CSF) examination were normal. Two days after admission he developed a right hemiparesis, and a brain MRIs showed bilateral cortical hyperintense signal on T2 sequences with leptomeningeal enhancement (Figure 2). After steroid initiation (methylprednisolone 40 mg/day) a remarkable neurological improvement was noticed. Extensive serological and liquoric evaluations were performed without significant findings. The dramatic and immediate patient’s response to steroid and MRI data strongly suggested a disimmune etiology. Over the ensuing week, his language, motor, and sensory function continued to improve. Two weeks after admission, the patient was discharged to home without significant neurologic sequela.

DISCUSSION Neurologic involvement in the course of Scleromyxedema represents a potentially fatal complication without a widely accepted pathological mechanism. Despite rare, a variety of central nervous system and neuromuscular conditions have been described, such as behavioral abnormalities, encephalopathy, seizures, cerebrovascular insults, peripheral nerve disorders, and myopathy. Mucin deposition is insufficient to explain all the systemic manifestations of scleromyxedema. It has been hypothesized that paraproteinemia can potentially explain transient focal neurologic disturbances, mainly through hemodynamic mechanisms. However a disimmune etiology should be considered as well, as our patient’s history is similar to that of other patients with scleromyxedema suffering from stroke or a stroke-like disorder. There are no consensual data on treatment of neurological complications of scleromyxedema; plasmapheresis, immunosuppressive therapy, intravenous immunoglobulin as well as high-dose steroids [3] have been reported. In summary, neurological dysfunction associated with scleromyxedema is a rare, possibly fatal and challenging entity. The recognition of this potentially treatable condition is based on the characteristic skin lesions.

REFERENCES