Scleromyxedema (papular mucinosis) with dermato-neuro syndrome: A rare, potentially fatal complication

Scléromyxédème (mucinose scléro-papuleuse) avec syndrome dermato-neurologique : une complication rare, potentiellement fatale

Scleromyxedema, or papular mucinosis, is a rare dermatological disease characterized by an accumulation of mucin in the dermis of patients with fibroblast proliferation and fibrosis, associated with a serum benign monoclonal gammopathy. Systemic manifestations have been described. Neurologic involvement includes myopathies, carpal tunnel syndrome, peripherical neuropathies, and encephalopathies with grave prognosis.

We present the fatal case of a man with recurrent episodes of encephalopathy with seizures, impaired consciousness and coma, and a flu-like prodrome, named "dermato-neuro" syndrome.

Case report

This 56-year-old man was diagnosed on a skin biopsy with scleromyxedema (papular mucinosis) following the appearance 6 months before of a non-itching papular eruption extended to the whole body (see figures 1 and 2). This mucinosis was associated with a monoclonal immunoglobulin Ig G lambda on serum protein electrophoresis with immunofixation (7.3 g/l). On 01/12/2013, he complained of severe headache, and intense fatigue. On 01/16/2013, he presented a generalized seizure. He was admitted to the ER where he presented a coma and was transferred to the ICU for 4 days. A cerebral CT scan was normal. Laboratory tests were normal apart from elevated...
creatine kinase (CK) at 1868 IU/L (39–308). EEG was normal, and brain MRI found nonspecific hyper-intensities.

On 10/03/2013, he presented an altered condition with a flu-like syndrome, which lasted for several days. On 10/09/2013, he presented an unexplained coma of sudden onset requiring intubation. He was hospitalized for 10 days in ICU and extubated with complete amnesia of facts. On 10/19/2013, he again presented a coma with respiratory distress requiring reintubation for 5 days. Blood work was within normal limits. Infectious and metabolic causes of encephalopathy were ruled out. CSF presented a coma with respiratory distress requiring reintubation. He was hospitalized for 10 days in ICU and extubated with normal glucose level, negative cultures. EEG found a discrete right occipital focalization. Treatment with levetiracetam was introduced, then. MRI features were unchanged.

On 06/30/2014, after a few days of flu-like episode, he was conducted to the ER for unconsciousness. He presented two generalized seizures followed by a coma requiring intubation. EEG performed showed no epileptic features but evoked an encephalopathy. CSF showed increased total protein level of 0.63 g/L (0.2–0.4) with normal glucose level, negative cultures. EEG found a discrete right occipital focalization. Treatment with levetiracetam was introduced, then. MRI features were unchanged.

On 10/09/2013, he presented an altered condition with a flu-like prodrome. He presented two generalized seizures followed by a coma requiring intubation. EEG performed showed no epileptic features but evoked an encephalopathy. CSF showed increased protein level of 2.3 g/L without any white cell and cultures were negative. He died 3 days later.

Discussion
Diagnosis criteria of the classification of scleromyxedema proposed by Rongioletti and Rebora [1] include:

• generalized popular and dermato-neuro syndrome;
• skin histological examination revealing mucin deposition in the reticular dermis, fibroblast proliferation and fibrosis;
• monoclonal gammopathy;
• the absence of thyroid disease.

Systemic, even lethal manifestations are common. Cardiovascular, pulmonary, renal, and rheumatologic manifestations have been described. Encephalopathies with grave prognosis appear not uncommonly during scleromyxedema without their frequency being accurately known [2]: central nervous system impairment is found in 10% of patients with scleromyxedema by Rey et al., [3]; confusion, coma, psychiatric disorders and focal neurological deficit can be observed [3]. A syndrome, sometimes fatal, consisting in fever, seizures and coma with a flu-like prodrome can rarely occur and is named “dermato-neuro syndrome” [4,5]: Our patient had this particular presentation.

Several assumptions have been made regarding the pathophysiology of neurological impairment in scleromyxedema [2]: damage secondary to the monoclonal gammopathy with hyperviscosity, mucin deposition in the brain. An autopsy of a patient with scleromyxedema and central neurological, gastrointestinal, and cardiac muscle involvement, demonstrated mucin only in the dermis, the coronary vessels, and in the lung [6].

Cases of remission have been described in the literature with different treatments: thalidomide [7,8], corticosteroids [9], plasmapheresis [10]. High-dose intravenous immunoglobulins [3,9] seem to have the best efficiency, even in cases of dermato-neuro syndromes. Considering the potentially fatal outcome of this disease, these treatments deserve to be better known.

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References

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