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

Scléromyxœdème (mucinose scléropapuleuse) 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 study showed increased total protein level of 0.63 g/L (0.2–0.4)

Figures 1 and 2

Multiple wide-spread papular eruptions measuring 2 mm on the back, arms, neck and trunk of the patient.
Un abcès hépatique secondaire à la migration d'un corps étranger ingéré

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Liver abscess caused by migration of an ingested foreign body

References


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