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, peripheral 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

Liver abscess caused by migration of an ingested foreign body

Anne F. Landais1, Célina M. Duchemin2, Véronique M. Bourhis2
1CHU de Pointe-à-Pitre, university hospital of Pointe-à-Pitre, neurology department, route de Chauvel, 97739 Abymes, Guadeloupe
2CHU de Pointe-à-Pitre, university hospital of Pointe-à-Pitre, oncology department, route de Chauvel, 97739 Abymes, Guadeloupe

Correspondence: Anne F. Landais, CHU de Pointe-à-Pitre, University Hospital of Pointe-à-Pitre, Neurology department, route de Chauvel, 97739 Abymes, Guadeloupe
landais-anne@voila.fr

Received 14 March 2015
Accepted 10 April 2015
Available online: 4 June 2015


avec un glucose sanguin normal, cultures négatives. L’EEG a montré une activité épileptiforme focale droite. Le traitement par levetiracetam a été introduit. Les signes de détresse neuronale ont été persistants. La voie respiratoire a été assurée par intubation. Les patients sont restés inconscients pendant 3 jours.

Discussion

On a discuté et examiné les critères de diagnostic et les termes proposés par Rongioletti et Rebora [1].

Conclusion

Le cas de l’abcès hépatique secondaire à la migration d’un corps étranger ingéré est rare. Son diagnostic est essentiel pour le traitement approprié.

References