A rare case of scleromyxedema complicated by neurological manifestations

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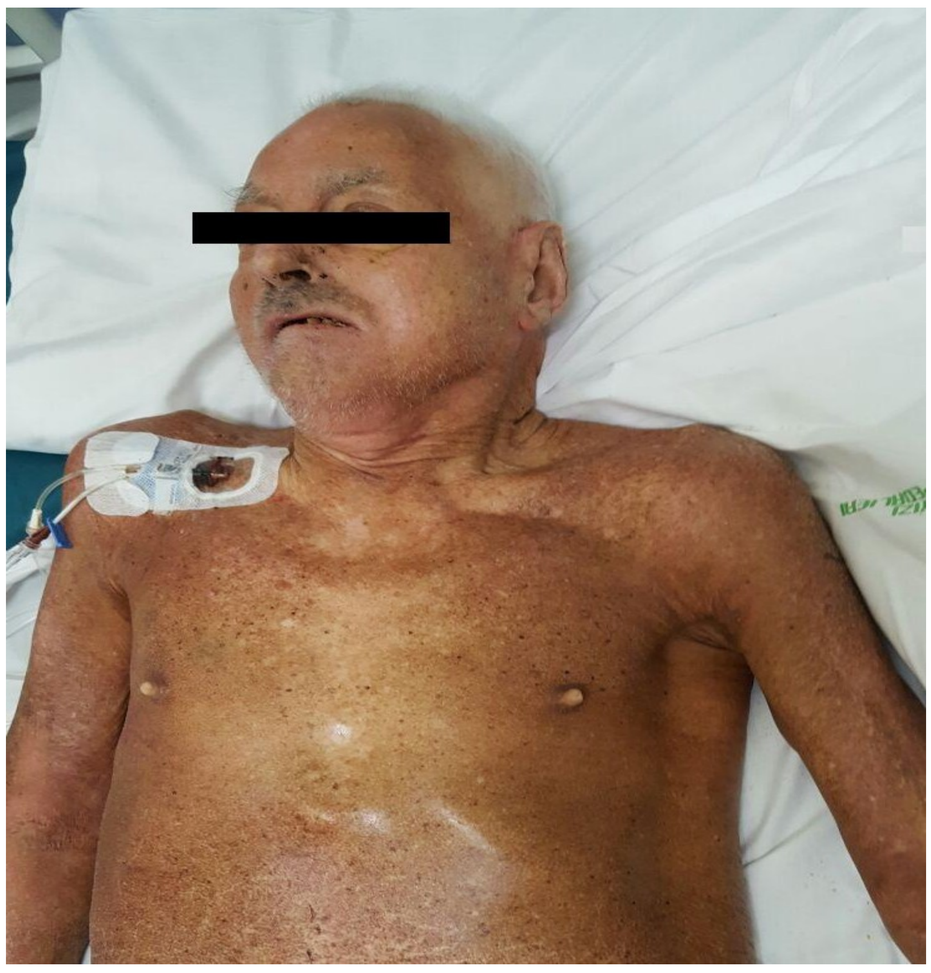

Fig.3 Diffuse induration , thickening and hyperpigmentation of the skin . Excessive thickening of the skin on the face, especially surrounding the lips, led to facial expression impairment..

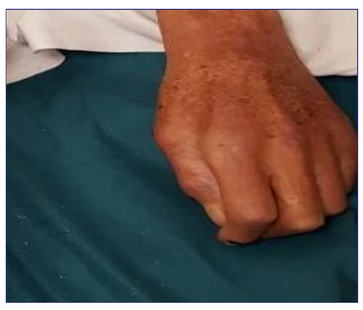

Fig. 4 Sclerodactyly due to induration, tightness, and infiltration of the skin.

Case Report:
In June 2016 a 76-years old man was hospitalized for brief episodes of right hemisoma hypoesthesia associated with transient language impairment, interpreted as transient ischemic attack. Medical history included scleromyxedema treated with periodic cycles of plasmapheresis and intravenous immunoglobin. In October 2016, after few days of flu-like syndrome, he was admitted to the Emergency Room for fever, confusion and disorientation. After few hours he developed tonic-clonic generalized seizures. Physical examination revealed an axillary temperature of 39°C, blood pressure of 145/80 mm Hg, pulse rate at 133 beats per minute. Computed tomography scan and electroencephalogram were normal. Magnetic resonance imaging was normal except for basal ganglia high magnetic-susceptibility substance accumulation (Fig.1) and pachymeningeal contrast enhancement related to lumbar puncture (Fig.2). The cerebrospinal fluid (CSF) revealed slightly elevated total protein (70 mg/dl, normal 18-43) and glucose (144 mg/dl, normal 74-106). Bacterial colture and polymerase chain reaction to detect herpes simplex virus in the CSF were negative. Serum protein electrophoresis and immunofixation showed IgG kappa light chain gammopathy. Patient was treated with antiepileptic drugs, Ceftriaxone and Acyclovir as encephalitis was suspected. Despite treatment, after 2 days he experienced generalized seizures uncontrolled by therapy, followed by coma with a Glasgow Coma Scale score of 7. All infectious, neoplastic and metabolic causes of encephalopathy were excluded, and due to his scleromyxedema history,a diagnosis of dermatoneuro syndrome was made. Treatment by intravenous immunoglobin at the dose of 2g/kg combined with methylprednisolone was started and patient regained consciousness and his general status slowly improved. He was discharged after 26 days with neurological sequelae, including dysphagia, dysarthria and motor disorder. After 7 days he was newly hospitalized because of heart attack and he died.

Discussion and conclusions:
Scleromyxedema is a rare disease characterized by a papular and sclerodermoid skin eruption due to a dermal mucin deposition[1](Fig.3-4) A monoclonal gammopathy is often present[1]. A rare but serious complication is dermatoneuro syndrome , consisting in fever, seizures and coma with a flu-like prodrome, which can have a mortality rate of 21%[2]. The mechanism responsible for Dermatoneuro syndrome remains unknown[3]. The cause of death of our patient seems to suffrage the hypothesis that an increase in blood viscosity, or the formation of neutrophil aggregates, both determined by paraproteinaemia[3], alter microcirculation and are responsible of cardiac ischemia and encephalopathy. Our purpose is to focus on this often misdiagnosed condition. The development of focal neurological signs in patients with scleromyxedema seems to be a negative prognostic factor, and it is mandatory a multidisciplinary approach to early recognize and treat in order to reduce its morbidity and mortality.

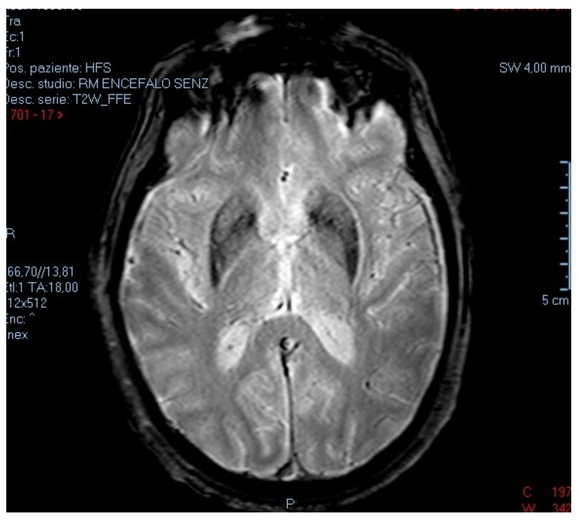

Fig.1 Magnetic resonance imaging demonstrating basal ganglia high magnetic-susceptibility substance accumulation

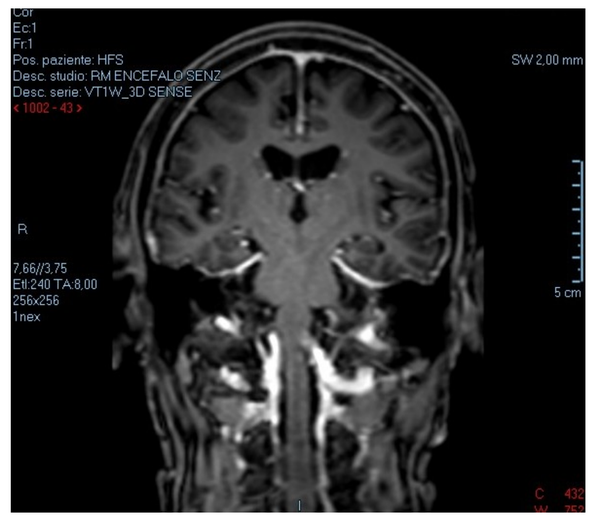

Fig.2 Contrast T1 coronal sequences showing pachymeningeal enhancement related to lumbar puncture

References
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