Clinical features of scleromyxoedema in an Afro-Caribbean man

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Accepted 4 December 2018

DESCRIPTION

Scleromyxoedema can be a challenging diagnosis on clinical examination with Afro-Caribbean skin.

We report on a case of a 61-year-old Afro-Caribbean man who presented to a public dermatology clinic with a greater than 30-year history of diffuse facial papules that had been previously treated as rosacea. His other complaints included tingling of the fingertips and enlargement of the ears. His medical history was pertinent for stroke, myocardial infarction, renal insufficiency, abdominal aortic aneurysm and hypertension. On examination, he had firm, skin-coloured monomorphic papules on the face, back, chest, arms and hands. There were firm, skin-coloured nodules on the central face, ears and glabella. Deep furrows on the forehead and glabella resulted in leonine facies (figure 1). His ears had a cauliflower appearance with prominent, firm cystic nodules involving the helix and lobes (figure 2). His torso had deep longitudinal furrows resulting in folds of hyperpigmented, indurated skin (figure 3). His hands had restriction in range of motion with hyperpigmented, annular nodules most prominently on the metacarpophalangeal joints (figure 4). An arrhythmia was detected on physical exam. There was no prior exposure to gadolinium.

Figure 1 Diffuse skin-coloured papulonodular cysts with greater predominance on the nose. Thickening of the dermis on the forehead and glabella resulted in leonine facies.

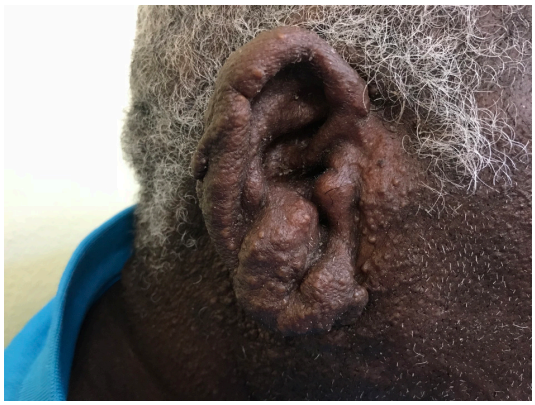

Figure 2 Firm nodules on the helix on the lobe of the ear produced a cauliflower appearance.

Laboratory examination revealed monoclonal gammopathy on serum protein electrophoresis (gamma-globulin, 2.5 g/dL (0.5–1.6 g/dL)) and normal thyroid function tests. The clinical characteristic of the facial lesions resembled cysts; however, histopathology showed increased dermal mucin confirming scleromyxoedema. His treatment consisted of 2 g/kg intravenous immunoglobulin (IVIg) 4 years prior to this presentation and numerous surgical procedures to debulk the lesions. He was lost to follow-up for several years and re-presented after a recent admission for a stroke.

Scleromyxoedema is a rare generalised papulonodular skin condition that is poorly understood. It is characterised by proliferation of fibroblasts and depositions of mucin within the dermis.[1]

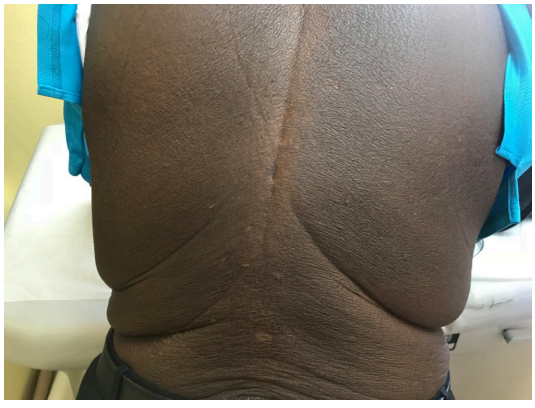

Figure 3 Deep longitudinal furrows of the torso causing folds of indurated skin.

To cite: Ufodiama CE, Tynes G, Unwala R. *BMJ Case Rep* 2019;**12**:e227588. doi:10.1136/bcr-2018-227588

Figure 4 Bilateral hyperpigmented nodules on the hands, predominantly around the metacarpophalangeal joints.

Its aetiology is unknown but has been associated with monoclonal paraproteinaemia and extracutaneous manifestations. The disease is progressive and chronic and has been reported to cause neurological, cardiac, rheumatologic, gastrointestinal and pulmonary complications; in particular myocardial infarctions and cerebrovascular accidents, this thought to be due to increased risk of atherosclerotic plaque development.[1 2] Plasmacytosis is commonly reported; it is possible for this to progress to multiple myeloma, but reports are, so far, rare. The diagnosis and prognosis of scleromyxoedema has been made easier through the development of a classification and diagnostic criteria.[1 2]

Treatment options vary, including the use of thalidomide, glucocorticoids, plasmapheresis and stem cell transplants.[3] IVIg and melphalan have been reported to have the best response; however, melphalan has been associated with haematological malignancies and sepsis, while IVIg is known to increase risk of thromboembolic episodes and there has been a report of cardiac toxicity.[2 3] Long-term use of IVIg as a form of maintenance therapy reduces rate of progression of the disease.[2]

Learning points

- A diagnosis of scleromyxoedema should be considered in patients with a history of recurrent facial cysts and leonine facies or skin thickening.
- Patients with scleromyxoedema should have regular surveillance for systemic involvement including neurological, rheumatologic and cardiovascular complications.
- Long-term maintenance therapy with intravenous immunoglobulin may delay progression of the disease.

Contributors CEU: conception, literature review, drafting of the article, final approval of published article. GT: critical revision of draft, final approval of published draft. RU: conception, acquisition of photos, critical revision of draft, final approval of published draft.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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