SKIN MANIFESTATIONS OF INTERNAL DISEASE

A CHALLENGING CASE OF ROSAI-DORFMAN DISEASE WITH IGG4

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Background: Rosai-Dorfman Disease (RDD) is a rare non-Langerhans cell histiocytosis associated with accumulation of activated histiocytes within the affected cells. It was first described in 1965 by Pierre Paul Louis Lucien Destombes, a French pathologist. RDD is a heterogeneous entity that can occur as an isolated disorder or in association with autoimmune, hereditary, and malignant diseases. Whilst most cases of RDD can be treated conservatively with close monitoring or skin directed therapies, a minority of RDD cases require systemic treatment for multifocal disease with associated morbidity and mortality.

Observation: A 47 years old Afro-Caribbean lady presented with a 6month history of multiple asymptomatic indurated, erythematous papules, nodules and plaques on her thighs bilaterally. A deep incisional skin biopsy showed features in keeping with RDD. Screening CT and MRI scans revealed retroperitoneal and sacral fibrosis associated with retroperitoneal and para-aortic lymphadenopathy. A retroperitoneal Lymph node biopsy excluded an underlying lymphoma. Despite treatment with super potent topical steroids, Protopic ointment, Dapsone, Azathioprine and Mycophenolate Mofetil, the patient's cutaneous lesions and retroperitoneal fibrosis continued to progress. The patient has now completed her first course of Rituximab and is awaiting follow up to assess for remission.

Key message: We have presented this case as an important reminder of the diagnostic, therapeutic and prognostic challenges associated with RDD. Multidisciplinary collaboration is often vital to the evaluation and management of patients with RDD, and systematic investigation of novel therapies for RDD is needed. The mechanism of action of Rituximab, an anti-CD20 monoclonal antibody is not entirely clear, but it is thought to improve the antibody-mediated pathogenetic mechanism involved in the disorder. The efficacy of rituximab has only been described in a few cases (especially in autoimmune-related RDD), although refractoriness and recurrences have also been described. Further studies evaluating its efficacy are required.