(***Refer to page 89***)

ANSWER: JUVENILE DERMATOMYOSITIS (JDM) WITH SOFT TISSUE CALCINOSIS

Juvenile dermatomyositis (JDM) is a rare autoimmune inflammatory disease involving the skin and striated muscles. The peak incidence is from 5 to 14 years of age, with variable female:male ratios depending on geographical distribution. It is estimated that 30-70% of children with JDM will have calcinosis.[1,2]

Clinical features of JDM include characteristic skin rashes (heliotrope rash, Shawl's sign, Gottron's papule) and symmetrical proximal muscle weakness. Muscle enzymes (CK, LDH, AST, ALT, aldolase) may be elevated. [3] Electromyography will show changes compatible with myopathy and muscle biopsy may show necrosis and inflammation, although both of these investigations are rarely used these days.[1] The Bohan and Peter criteria and the newer European League Against Rheumatism and American College Rheumatology (EULAR/ACR) inflammatory idiopathic myositis (IIM) classification criteria are used for diagnosis.[3]

Calcification is usually seen at the site of necrosis, in the chronic phase of the disease and may affect the skin, subcutaneous tissue, fascial planes and muscles. Four different patterns of calcification are recognized. These lead to more disability and limitations than myopathy alone. Superficial calcific masses can lead to discharging sinuses and non-healing ulcers.[2]

On plain radiograph, JDM typically shows calcification in the muscles and soft tissues (calcinosis universalis), classically seen affecting the thigh muscles. MRI T2-weighted sequences has been used to assess the affected muscles and calcified areas and to determine the disease activity and as a guide for muscle biopsy.[3]

Treatment for JDM includes high dose corticosteroid initially with disease-modifying drugs like methotrexate or cyclosporin A. Early and aggressive treatment may prevent complications like calcinosis. Various medications like bisphosphonates, calcium channel blockers and high dose steroids have been used for treatment of calcinosis.[1,2]

The present patient is under the joint care of dermatology and rheumatology since the diagnosis of JDM in 2015. He has been treated with methotrexate and prednisolone. EMG showed changes of myopathy and typical MRI changes have also been noted in the thigh muscles.

REFERENCES

1: Martin N, Li CK, Wedderburn LR. Juvenile dermatomyositis: new insights and new treatment strategies. Ther Adv Musculoskel Dis. 2012;4:41-50. doi:10.1177/1759720X11424460. [Accessed on the 22nd June 2019]. Pdf available at https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3383527/pdf/10.1177_1759720X11424460.pdf

2: Sanyal S, Atwal SS, Mondal D, Garga UC: Radiograophic Patterns of Soft Tissue Calcinosis in Juvenile Dermatomyositis and its Clinical Implications. J Clin Diagn Res. 2014;8:RD08–RD11. doi: 10.7860/JCDR/2014/10787.5321. [Accessed on 22nd June 2019]. Pdf available at https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4316317/pdf/jcdr-8-RD08.pdf

3: Lundberg IE, Tjärnlund A, Bottai M, et al.: 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups. Annals of the Rheumatic Diseases 2017;76:1955-1964.