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FIBROBLASTIC RHEUMATISM: A CASE REPORT

Camilla de Castro e Silva (UNIVERSIDADE DO ESTADO DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil), Camila Pitasi Arguelhes (UNIVERSIDADE DO ESTADO DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil), Victor RebelLo Vertzman (UNIVERSIDADE DO ESTADO DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil), Pedro Felipe de Almeida Vianna (UNIVERSIDADE ESTADUAL DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil), LAISSA CRISTINA ALVES ALVINO (UNIVERSIDADE ESTADUAL DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil), Talita Gonçalves Pereira de Souza (UNIVERSIDADE ESTADUAL DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil), Roger Abramino Levy (UNIVERSIDADE ESTADUAL DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil), Veronica Silva Vilela (UNIVERSIDADE ESTADUAL DO RIO DE JANEIRO, RIO DE JANEIRO, RJ, Brasil)

BACKGROUND

Fibroblastic rheumatism is a rare disease characterized by excess fibroblastic proliferation and increased collagen tissue deposition. It was described by Chaouat in 1980 and by now few cases have been reported. The clinical syndrome is composed of polyarthritis and cutaneous rapidly progressive fibrous nodules resulting in irreversible skin and joint contractures. The differential diagnosis includes lymphocytic reticulohystiocitosis, rheumatoid arthritis and systemic sclerosis. There is no effective treatment. Several drugs, including imunossupressants, colchicine, D-penicilamine and anti-TNF were used with scarce reports of improvements

CASE REPORT

A 43 years old female patient with a presented with hand palmar violaceous nodules. She had a past 10-years diagnosis of Sjogren´s syndrome in treatment with hydroxicloroquine. The initial skin biopsy conclusion was of palmar mucinosis. After six months, the nodules rapidly increased and spread to the hands, forearms, foots and legs, resulting in severe contractures, mainly on the hands. She loss 8kg and became disabled. Concurrently, she had hand and ankles arthritis, positive ANA, anti-Ro and polyclonal hypergamaglobulinemia and a nailfold capillaroscopy disclosing scleroderma LIKE pattern. Right hand magnetic resonance disclosed joint effusions with reactional osteitis and heterogeneous thickening of deep flexors tendons. A new hand skin biopsy disclosed lymphocytic perivasculitis, tenosynovitis with hyaline fibrosis, proliferation of fusiform and cells and muscle atrophy. A diagnosis of fibroblastic rheumatism was concluded. Methotrexate 25mg/week was given. Hand surgery was performed to release the tendons and improve hand´s movements. Extensive investigation to exclude hidden neoplasy was negative. R-rays of the skeleton did not reveal plasmacytic appearance. After six months of methotrexate, the patient did not improve. A short period of etanercept also was not effective. After one year, the fibrosis progressed and the patient remains disabled.

CONCLUSION

Fibroblastic rheumatism is a rare disease that results in skin and joints infiltration with fibrosing nodules. The diagnosis is based on clinical features and histologic confirmation. It can be considered as a scleroderma mimicker. Exclusion of paraneoplastic syndrome and lymphocytic reticulocitosis is mandatory. We here describe the case of a female patient with a severe form of the disease. She previously had Sjogren´s disease, which could corroborate some link with an autoimmune background. We emphasize the need for the extensive investigation to exclude malignancy or other overlap systemic autoimmune disease. The dismal prognosis presented in this case should make us aware that more research for treatments for fibroblastic rheumatism is necessary.