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TOCILIZUMAB FOR THE TREATMENT OF CORTICOID-RESISTANT ADULT-ONSET STILL'S DISEASE

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BACKGROUND

Adult-onset Still's Disease is a severe autoinflammatory condition traditionally characterized by the triad of arthralgia, persistent high fevers, and a characteristic salmon-colored skin rash. Furthermore, extremely elevated inflammatory markers ferritin values are generally noted. Since it is considered a diagnosis of exclusion, its investigation is often challenging, even in specialized centers.

CASE REPORT

A 44-year-old male started in March 2018 with arthritis of wrists, knees, hands and elbows, evolving to symmetrical polyarthralgia. He denied prolonged morning stiffness. However, he mentioned important fatigue which limited his daily activities. He was originally treated by a private practice rheumatologist, who was initially diagnosed him with rheumatoid arthritis and started treatment with Methotrexate 12.5 mg/week; Hydroxychloroquine 400mg q.d. and Naproxen 500mg bid with significant improvement. In September 2018, the patient developed erythematous and pruritic lesion in his right thigh, later progressing to new papulomacular lesions in the trunk and limbs, as well as daily fevers around 39º. The subject was admitted at another service with the suspected diagnosis of an infection without focus. Antibiotic regimens with Cefotaxime and Oxacillin were unsuccessful. After prolonged hospitalization without improvement, the patient was admitted to our service for investigation. He persisted with arthritis, fever and a skin rash. Initial laboratory workup presented significant leukocytosis of 41,840/mm^3 with 31% rods, C-reactive protein of 15.6 mg/dL, ESR of 24 mm and ferritin of 147,451 ng/mL. After eliminating any possible infectious conditions, the diagnosis of Adult-onset Still's Disease was made and the treatment initiated with an 1g pulse of intravenous methylprednisolone and maintenance dose of 1mg/kg /day. The patient, however, persisted with fever and arthralgia, despite treatment. Due to the unavailability of IL-1 antagonists (Anakinra), which are the first line of treatment in case of corticoid resistant disease, it was opted for Tocilizumab, with a good response.

CONCLUSION

Adult-onset Still's Disease is a potentially serious condition and a diagnosis of exclusion, which can generate a plethora of difficulties in its investigation. Our case presented a patient with corticoid resistant disease and Tocilizumab was a satisfactory alternative to Anakinra.