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Fever of Unknown Origin: Adult Onset Stills Disease

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I. Introduction

Adult onset still diseases (AOSD) is an uncommon clinical entity that predominantly effects young adults. Usually present as fever of unknown origin. It is a multi systemic inflammatory disorder of unknown etiology characterized by spiking fever, skin rash, arthralgia/arthritis and myalgia.

II. Case Report

A 28 year old male presented with complaints of high grade fever, joint pain, muscular pain & dry cough since 9 days. He had no significant medical or drug history. Marked pain was present in all joints and also thigh muscles. Physical examination showed petechial hemorrhagic spots on palate. The abdominal examinations showed mild spleenomegaly, other systemic examination was normal. TLC (11000), ESR (80) were elevated, CRP+, ASLO titer <200. ANA and RA factor negative. Sr. ferritin level value was > 40,000 mg/ml.

X-Ray showed B/L minimal pleural effusion, MP card and widal was negative. Patient was given symptomatic treatment but was not relieved.

As per yamaguchi criteria patient full filled 6 criteria including 4 major criteria and 2 minor criteria and was diagnosed to be case of AOSD. Patient was started on prednisolone 1mg/kg body weight and methotrexate 10mg/week. Patient showed marked improvement after initiation of treatment.

III. Discussion

Adult onset Still's disease (AOSD) is a rare inflammatory disorder that affects the entire body (systemic disease). The cause of the disorder is unknown (idiopathic). Affected individuals may develop episodes of high, spiking fevers, a pink or salmon colored rash, joint pain, muscle pain, a sore throat and other symptoms associated with systemic inflammatory disease. The specific symptoms and frequency of episodes vary from one person to another and the progression of the disorder is difficult to predict. In some individuals, the disorder appears suddenly, disappears almost as quickly and may not return. In other people, adult onset Still's disease is a chronic, potentially disabling, condition. Various medications are used to treat individuals with adult onset Still's disease, affected individuals may respond to therapy differently. Adult onset Still's disease does not appear to run in families.

Adult onset Still's disease is the adult form of systemic juvenile rheumatoid arthritis (juvenile Still's disease). The disorders are name after a British physician who first described systemic juvenile rheumatoid arthritis in the medical literature in 1896. The term "adult Still's disease" was first used in the medial literature in 1971, but cases that fit the description of the disorder appear in the medical literature as early as the late 1800s.

IV. Conclusion

Adult-onset Still's Disease is rare and has been described all over the world. The cases of AOSD per year is estimated to be 0.16 new cases per 100,000 population. Prevalence is estimated at 1.5 cases per 100,000-1,000,000 population. Therefore patients with these presenting features should be worked upon well so that this rare disease can be treated well.