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Beyond the Usual Suspects: Unraveling a Case of Kikuchi-Fujimoto Disease

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INTRODUCTION

A case of Kikuchi-Fujimoto Disease (KFD); a rare disorder with clinical presentation mimicking other diseases in a young boy.

CASE REPORT

A healthy 15-year-old teen, presented with unremitting fever for 1 week associated with myalgia, arthralgia, and unintentional significant weight loss. He also noticed rashes distributed from trunk down to his limbs since day 1 of illness. Otherwise, he reported no infective symptoms, autoimmune symptoms nor any significant family history of malignancy or recent travel history. He also denied taking any illicit drugs. Besides, he did not recall having any tuberculosis contact, sick contact or B symptoms prior or during the index event. On day 8 of illness, he was brought into the Emergency Department with the main concern of unresolved fever. Clinically, he was hemodynamically stable. There were diffuse, palpable lymph nodes over right inguinal and left cervical region tethered to the skin. Also, there was maculopapular rash over the trunk and all 4 limbs with splenomegaly upon abdomen palpation. Initial lab investigations show hyperleukocytosis. A provisional diagnosis of lymphoma with differential diagnosis of occult sepsis were made. A dermatology review commented as maculopapular exanthem secondary to underlying infection. A myriad course of antibiotics was commenced empirically. Meanwhile, extensive investigations were performed which included full septic workup and also screening for both autoimmune condition and occult viral infection. However, all turned out to be negative. In view of the background epidemiology of Sabah, tuberculosis workups were done including a chest X-ray, three gastric lavage acid AFB stain and a gastric lavage tuberculosis gene expert stain which resulted as negative. We performed a computer tomography scan of the thorax, abdomen and pelvis which reported as splenomegaly with axillary, mediastinal and inguinal lymphadenopathy each measuring about 1-1.5cm (Figure 1). Hence, lymph node excisional biopsy was performed and HPE shown to be histiocytic necrotizing lymphadenitis with residual lymphoid follicles and germinal centers. Diagnosis of KFD was made and symptomatic treatment with a short course of corticosteroid was started with intravenous hydrocortisone for 3 days which followed by oral prednisolone 0.5mg/kg upon discharge.

CONCLUSION

Clinician tend to face diagnostic challenges during the course of investigating a rare disease. The importance of acknowledging such a rare disease is highlighted in this case.

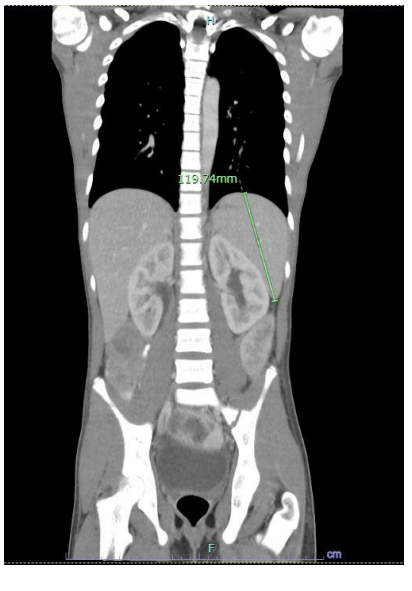

Figure 1: Contrast CT Thorax, Abdomen and Pelvis showing splenomegaly measuring 11.9cm

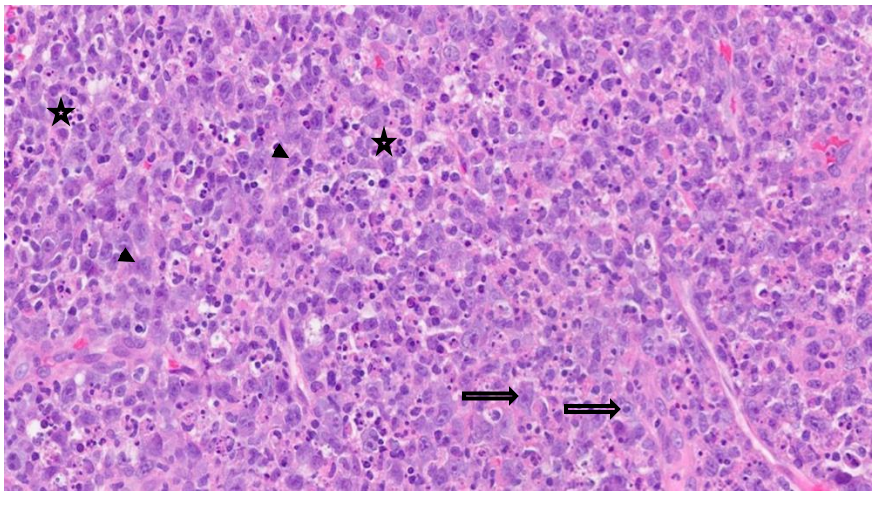

Figure 2 : Image shows karyorrhetic debris (star) mixed with proliferations of histiocytes (arrowhead), immunoblasts (arrow), lymphocytes and plasmacytoid dendritic cells.