Hemophagocytic Lymphohistiocytosis (HLH) with History of Kawasaki Disease (KD)

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Hemophagocytic Lymphohistiocytosis (HLH) is an aggressive inflammatory systemic disease characterized by hyperinflammation and dysregulation of the immune system. HLH can be secondary to infections, such as Epstein-Barr virus (EBV), malignancy, immunodeficiency and Kawasaki Disease (KD) or primary due to familial mutations. Clinical manifestations are not specific, such as fever, skin rash, lymphadenopathy.

KD is a form of vasculitis affecting the medium-sized vessels; it is diagnosed by clinical criteria as there is no laboratory test to confirm the diagnosis. Clinical manifestations include fever, skin rash and lymphadenopathy. There is overlap of clinical manifestations of HLH and KD.

We report a three-year-old female who presented with clinical features of KD, but further investigations revealed the diagnosis of HLH. HLH is a rare disease and presents with non-specific clinical manifestations which overlap with other diseases. The diagnosis is often challenging and delayed.

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