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Title: Regression of porto-pulmonary hypertension in a iMCD patient under siltuximab

Abstract:

Background:

Pulmonary hypertension is an uncommon complication of idiopathic multicentric Castleman's Disease (CD, iMCD) that, in the Japanese cohort, has been reported to respond to tocilizumab. Portal hypertension due to nodular regenerative hyperplasia of the liver (NRH), secondary to iMCD, is a rarer complication, and data are lacking regarding response to IL-6 antagonism.

Case Presentation:

We describe a male who presented at age 32 with a mediastinal mass with a pathology in the CD spectrum, and no autoinflammatory symptoms. Two years later, he developed progressive lymphadenopathy, clinical and biochemical hyperinflammation, and liver and heart failure; he had evidence of portal hypertension with esophageal varices, hypertensive gastropathy and splenomegaly, with liver biopsies revealing diffuse NRH. Concurrent pulmonary hypertension was diagnosed by echocardiography and catheterization.

He was treated with 6 cycles of CHOP, with imaging, clinical and biochemical responses, but persistent porto-pulmonary hypertension and NYHA Class III heart failure, despite sildenafil.

He remained in iMCD remission over the following 9 years. In November 2019, he presented with progressive iMCD, and started siltuximab in January 2020. He was a rapid responder, with a biochemical complete response within a week of the first infusion, and a remission of his hyperinflammatory symptoms. Simultaneously, porto-pulmonary hypertension, liver failure and heart failure have progressively regressed, with a gradual decrease in medication needs. By May 2023, the patient had been restaged as NYHA Class I, had returned to work full-time, and went on a 25-mile hike with no complications. As of November 2023, he remains well, under treatment."