Regression of porto-pulmonary hypertension in an iMCD patient under siltuximab

INTRODUCTION

Pulmonary hypertension
• Uncommon complication of idiopathic multicentric Castleman’s Disease (iMCD)
• Reported to respond to tocilizumab in the Japanese cohort
Portal hypertension
• Due to nodular regenerative hyperplasia of the liver
• Rarer complication of iMCD
• Data are lacking regarding response to IL-6/IL-6R antagonism

CASE PRESENTATION

Benjamin Castleman
Describes “Castleman’s disease”

1956
♂ patient
Born in 1975

1975
Tocilizumab approval (Japan)

2005
32 years old
• Mild congenital factor V deficiency
• Baseline activity: 25-40%
• No other relevant clinical history

2007
• Mediastinal lymphadenopathy mass
• Supravacular lymphadenopathies
• No autoinflammatory symptoms

Surgical excision (November)
Pathology in the CD spectrum (hyaline-vascular)

2009
34 years old
Progressive supradiaphragmatic lymphadenopathy

Surgical excision (May)
Pathology in the CD spectrum (hyaline-vascular)

HHV-8 latent nuclear antigen: negative
MIV 1/2: negative

1975
PORTO SECTOR, A (latent nuclear
Moderate to severe tricuspid regurgitation
Mild
Persistent heart failure
in the Japanese cohort

Disease
Severe enlargement of
Hypertension
Internal
decrease
PASP
Mild
Medicine
Progressive
C,
RCT
♂ negative

Castro
Asymptomatic
Clinical Hematology Department, Coimbra University Hospitals
needs
Sinusoidal distension
p
regression
heart
No
Due to nodular regenerative hyperplasia of the liver

CHUC)
Pulmonary hypertension
Hypercellular marrow
of
34
Class
(November)
35
W
Uncommon complication of idiopathic multicentric Castleman’s Disease

No
Hypertensive gastropathy
No
of
Graça
Gradual
Department,
heart
Department,
Rarer complication of
Rapid
Noncirrhotic portal hypertension
years old
I
(EMA approval
CHUC)
Stage
Covid
marta.dot
No other relevant clinical history
infection
to
on
C1D1
complications
(Pulmonary
hypertension
in
full
Returned
Splenomegaly
antagonism
active
b
(Hepatology
NYHA
Downstaged
a
Nuno
Maria
Complete clinical
regression
treatment
ongoing
or
Myeloid
2019
Department,
Went
megakaryocytic hyperplasia
Data are lacking regarding response to IL
medication
Esophageal
Persistent
failure

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