Multicentric Castleman Disease associated with Duchenne dystrophy and cutaneous leukocytoclastic vasculitis in a 4-year-old child

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Introduction

Idiopathic multicentric Castleman disease is a rare lymphoproliferative disease characterized by multifocal lymph nodes, severe inflammatory manifestations and overproduction of interleukin 6 (IL-6).

Clinical Case

A 4-year-old boy, with non-explored behavioral disorder, consulted for an abdominal distension, a poor general state and a low-grade fever for one month. He had been suffering from a drop in his weight-status curve for at least two years.

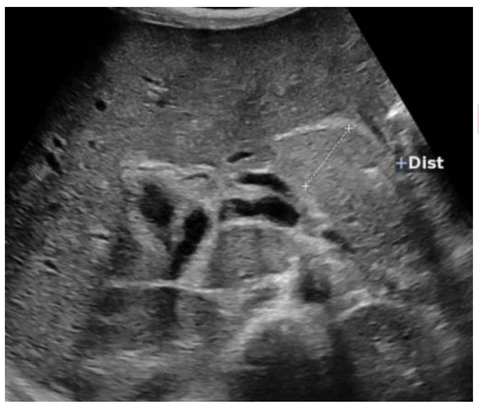
Clinically, he suffered from acute malnutrition, had a waddling walk, an incomprehensible language and a 6 cm hepatomegaly.

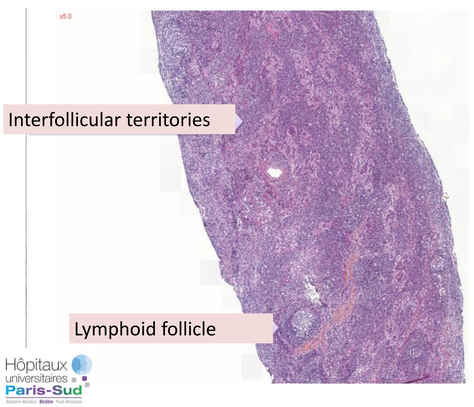
Lymph node biopsy showed specific characteristics of **Plasma-Cell type Castleman disease.**

Duchenne muscular dystrophy was diagnosed thanks to muscular biopsy and genetic analysis.

Hb	Platelets	CRP	ESR	Ig G/ Albu
10 g/dl	665000 /mm3	48 mg/l	131 mm	37/ 29 g/l

AST/ ALT/ CPK	Bili T/ Bili C	IL-6	Anti-smooth muscle and anti-actin Ab
7N/ 7N/ 2500 U/l	20/ 17 U/l	36 pg/ml	1/160[e]



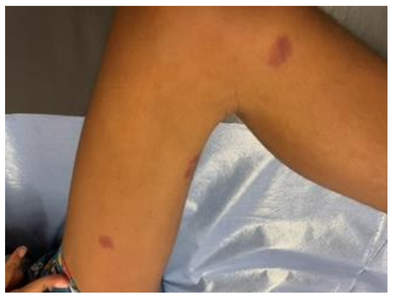


Abdominal imaging: Homogeneous hepatomegaly, adenopathies of the hepatic hilum, diffuse dilatation of the bile ducts with no identified area of compression

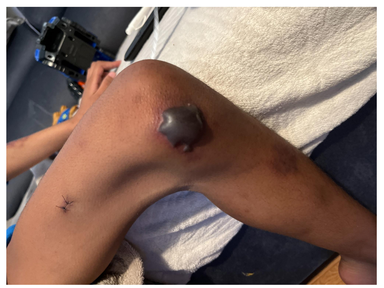
Hiistology of one of the adenopathies Preserved but altered architecture, atrophic lymphoid follicles with onion bulb outline of the mantle and hyperplasia of interfollicular territories with abundant plasma cell infiltrate.

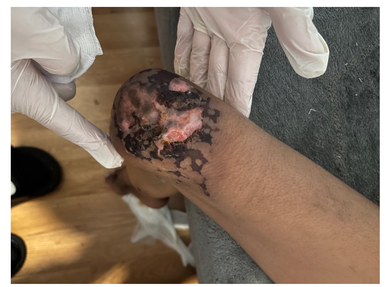
Treatment and evolution

The patient responded well to tocilizumab, an anti-IL-6 receptor monoclonal antibody. When tocilizumab infusions were spaced out, he developed episodes of cutaneous leukocytoclastic vasculitis with inflammatory digestive involvement. Tocilizumab infusions were brought closer together, every two weeks, and he is receiving enteral nutrition.



Polymorphic purpuric eruptions





Bullous skin lesions, and later necrotic and ulcerated lesions

Discussion

On initial presentation, muscle damage with CPK between 1300 and 20475 U/L, muscle weakness and cognitive impairment were not explained by Castleman disease. A muscle biopsy and molecular biology confirmed the diagnosis of Duchenne muscular dystrophy.

The patient also presented with episodes of highly polymorphic purpuric eruptions. The episodes have recently become more frequent and more severe, despite toclizumab infusions every two weeks. Biopsy of the lesions showed leukocytoclastic vasculitis with C3 and IgA deposits on immunofluorescence. The patient also presented concomitant episodes of arthralgia without true arthritis, and diarrhea. Treatment with colchicine has just begun and the episodes are treated with short courses of steroids.

Conclusion

This observation is the first to describe a boy with an iMCD associated with a Duchenne muscular dystrophy, an inflammatory disease of the digestive tract, and a cutaneous leukocytoclastic vasculitis.