Opis choroby *

Definicja

A rare systemic autoimmune disease characterized by mass-forming, potentially destructive inflammation and fibrosis in the soft tissues of the retroperitoneum, associated with elevation of serum IgG4 levels and infiltration of IgG4-positive plasma cells in at least one organ or site. Most frequent locations are peripheral to the abdominal aorta, as well as the iliac and renal arteries. Clinical symptoms are unspecific and include abdominal pain, back pain, and edema of the lower extremities. The condition may occur together with IgG4-related disease in other parts of the body.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
Idiopathic retroperitoneal fibrosis
Choroba Ormonda
Idiopatyczna włóknienie zaotrzewnowe
Ormond disease

Kod ORPHA
49041

Kod OMIM
228800

Kod ICD10
K66.2

Kod ICD11
4A43.0

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*Źródło

orphanet