

SENSORY AXONAL POLYNEUROPATHY IN A PATIENT WITH SYSTEMIC CAPILLARY LEAK SYNDROME (CLARKSON DISEASE): A CASE REPORT.

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Introduction

Systemic capillary leak syndrome, or Clarkson disease, is a very rare disease of unknown origin, first described in 1960 and reported overall in about 150 cases. It's characterized by unexplained episodic attacks of capillary leakage of plasma from the intravascular into the interstitial space, causing acute, recurrent episodes of hypotension, oedema and hypovolemia. Since its rarity, clinical features and comorbid conditions remain largely unclear. We describe the clinical case of a patient with a diagnosis of Systemic capillary leak syndrome, subsequently developing sensory axonal polyneuropathy.

Case Report

The patient's clinical history was unremarkable until the age of 48 years old, when he underwent a surgical procedure of duodenocephalopancreasectomy for a carcinoma of the ampulla of Vater, followed by 12 cycles of cisplatin and 5-Fluorouracil based chemotherapy, obtaining complete clinical remission.

At the age of 57 years old, the patient started to develop quite sudden episodes characterized by generalized weakness, fatigue and abdominal pain, rapidly followed, over few hours, by severe arterial hypotension, oliguria, oedema of the face and lower limbs and weight gain, requiring hospitalization and symptomatic management; the episodes lasted about 2-3 days and were followed by massive oedema resorption, leading to polyuria and weight loss.

Based on clinical history and laboratory findings of hemoconcentration and hypoalbuminemia during the episodes, in the absence of secondary causes of shock, the patient received a diagnosis of systemic capillary leak syndrome (Clarkson disease). Additionally, serum laboratory examinations revealed a monoclonal IgG gammopathy, without evidence of lymphoproliferative disorders.

At the age of 62 the patient progressively started to complain of distal paresthesias/dysesthesias at both upper and lower limbs; neurological examination showed mild ataxic gait, positive Romberg sign, deep and superficial sensory dysfunction of all extremities and diffuse osteo-tendon hyporeflexia. Electroneurographic examination provided evidence of sensory axonal polyneuropathy. Cerebrospinal fluid analysis showed mild albumin-cytologic dissociation. The patient initially performed oral corticosteroid therapy for about two months, with mild benefit on sensory disturbances, but unchanged electroneurographic findings.

Discussion

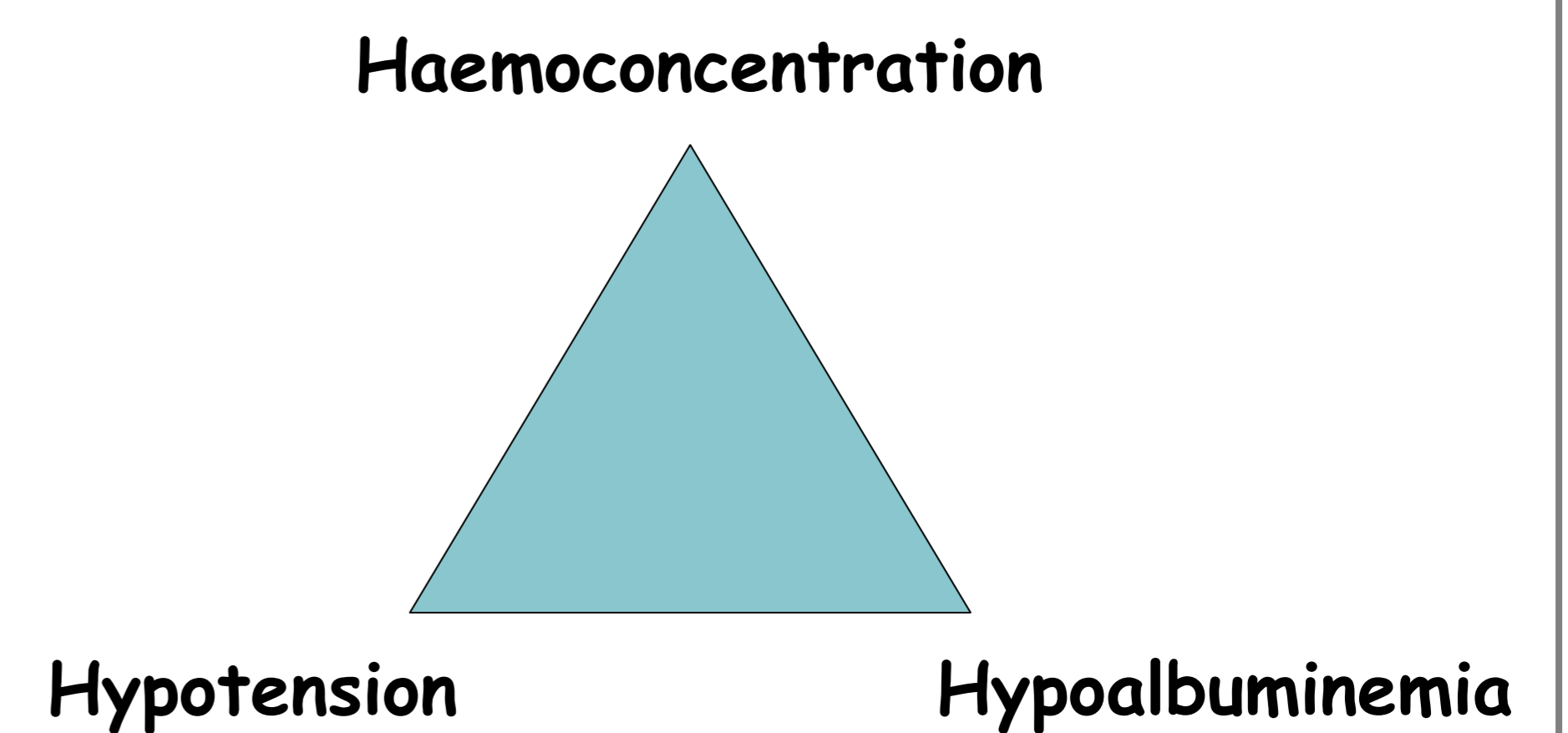
A case of sensory axonal polyneuropathy in a patient with Clarkson disease, IgG monoclonal gammopathy and previous chemotherapy for carcinoma of the ampulla of Vater is here reported.

Although the pathophysiological mechanisms leading to sensory axonal neuropathy in this complex case remain unknown, the present report seems to expand the spectrum of clinical presentation and comorbidities of Clarkson disease.

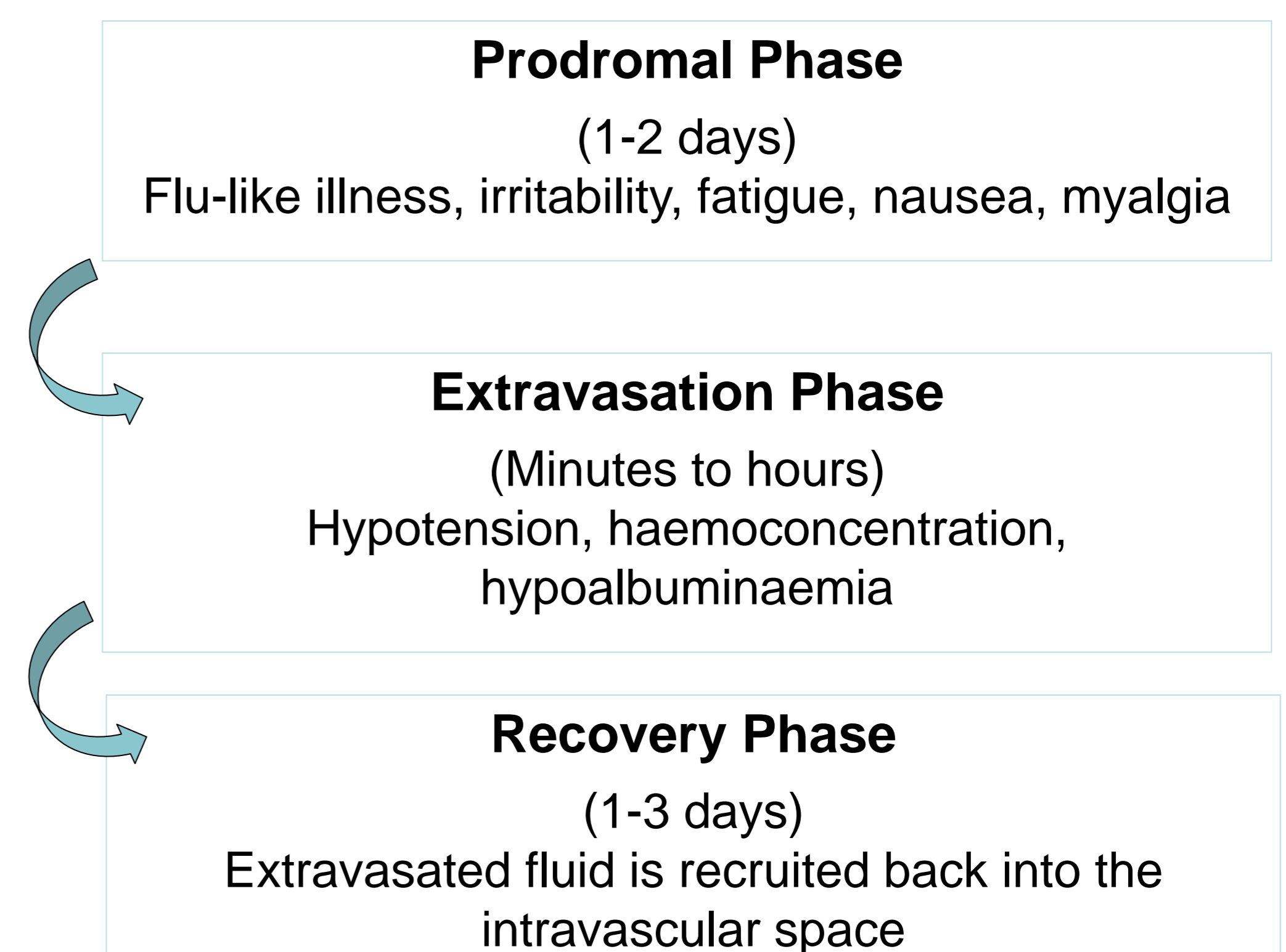
References

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CLASSIC TRIAD OF SYSTEMIC CAPILLARY LEAK SYNDROME



DISTINCT PHASES SEEN IN SYSTEMIC CAPILLARY LEAK SYNDROME



PATIENT'S CLINICAL PICTURE

Previous carcinoma of the ampulla of Vater (in remission, after surgical procedure followed by chemotherapy)

Systemic capillary leak syndrome (Clarkson disease)

Monoclonal IgG gammopathy (without evidence of lymphoproliferative disorders)

Sensory axonal polyneuropathy