中文題目：POEMS 症候群，以難治性腹水為一開始之表現--個案報告

英文題目：POEMS syndrome with initial presentation of refractory ascites - a case report

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Abstract:

Refractory ascites is defined as ascites that does not recede or that recurs shortly after therapeutic paracentesis, despite sodium restriction and diuretic treatment, and is commonly seen in patients with cirrhosis. POEMS syndrome is the acronym for polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, and skin changes. It is a rare disease caused by paraneoplastic syndrome of the plasma cell disorder. Most of patients with POEMS syndrome manifest with demyelinating polyneuropathy at the initial presentation. We hereby report the 46-year-old male who suffered from refractory ascites, 10 months before the diagnosis of POEMS syndrome. Although the analysis of his ascites showed a high serum-ascites albumin gradient, (1.92 g/mL) and suggested the portal-hypertension-related nature (only seen in 26% of POEMS syndrome), the clues to diagnose POEMS syndrome was the identification of IgA-λ monoclonal gammopathy and multiple endocrinopathies including hypothyroidism, adrenal insufficiency, hypogonadism, and growth hormone deficiency. Subsequent workup also found polyneuropathy, sclerotic bone lesion over left femur bone, splenomegaly, skin changes (hyperpigmentation and white nails), and papilledema. The ascites dramatically improved after the first cycle of chemotherapy with cyclophosphamide and steroid. There was further improvement of numbness of four limbs and anasarca after 3 cycles of therapy. To better control the disease, the patient will undergo high-dose chemotherapy and autologous peripheral blood stent cell transplantation. In conclusion, the POEMS syndrome must be added to the list of rare causes of refractory ascites. Although refractory ascites implied poor prognosis in POEMS syndrome, an early diagnosis and prompt therapy may improve the outcome.