Gastrointestinal involvement in POEMS syndrome: a novel clinical manifestation

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POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes) is characterised by a rare multisystem disorder of unknown pathogenesis. Although its pathophysiology is not well understood, overexpression of proinflammatory cytokines has been implicated. Gastrointestinal system disorders have not been reported among the components of the syndrome. A case is reported of POEMS syndrome with gastrointestinal involvement shown by gastrointestinal endoscopy.

CASE REPORT

A 37 year old woman was admitted to our hospital with three months’ history of polydipsia, polyuria, weight loss (8 kg in three months), dyspepsia, and diarrhoea (8–10 times/day), myalgia, bone pain, and progressive weakness of both lower limbs that left the patient chairbound. Her medical history was unremarkable. Physical examination showed mild hepatomegaly, axillary and inguinal lymphadenopathies, generalised skin hyperpigmentation, oedema, and moderate proximal and distal sensory loss. The remaining systemic examination was normal.

Laboratory examination showed a normal whole blood count and erythrocyte sedimentation rate. Serum transaminases, creatinine, and urea levels were within normal range. Serological testing for Helicobacter pylori was found to be negative. Stool leucocyte, ovum parasite evaluation, and also stool culture was negative. Although fasting plasma glucose level was 23.5 mmol/l (3.9–6.1), insulin therapy was started on admission. Test for rheumatoid factor, cryoglobulin, antinuclear antibody, anti-double stranded DNA antibody, and antineutrophil cytoplasmic autoantibody were negative. Bence Jones proteins were not detected. Electromyographic findings were consistent with demyelinating peripheral neuropathy. Computed tomograms of the abdomen showed multiple sclerotic bone lesions, hepatomegaly, and intra-abdominal lymphadenopathy with no splenomegaly. Portal Doppler ultrasonography was normal. Cranial magnetic resonance imaging showed decreased bone marrow intensity in all the skull bones suggesting myelofibrosis. Immune electrophoresis showed a monoclonal gammopathy (immunglobulin G lambda), and the serum IgG level was 15.81 g/l (7.51–15.60). Bone marrow histological examination showed diffuse atypical plasma cell infiltration. Inguinal lymph node biopsy showed angiofollicular hyperplasia with hyaline vascular transformation of the lymphoid follicles, which is characteristic for Castleman’s disease.

Gastric endoscopy showed a snakeskin-like appearance of stomach and duodenum (fig 1), and whole colonic mucosa was hyperaemic on colonoscopy. Multiple biopsy specimens from several regions of the colon mucosa were taken. Microscopic examination showed infiltration of lamina propria by numerous inflammatory cells, mainly by neutrophils (fig 2). Amyloid staining was negative.

A diagnosis of POEMS syndrome was reached based on the presence of polyneuropathy, hepatomegaly, endocrinopathy (diabetes mellitus), monoclonal gammopathy, and skin hyperpigmentation. The patient was treated with CHOP protocol including cyclophosphamide (1100 mg) for one day, adriamycin (75 mg) for one day, vincristine (2 mg) for one day, prednisolone (150 mg) for five days. After chemotherapy her myalgia, weakness, skin oedema, and paresis showed considerable improvement and also gastrointestinal symptoms including diarrhoea and dyspepsia disappeared. She was discharged 30 days after admission. A repeat upper gastrointestinal endoscopy and colonoscopy was performed 45 days after initial chemotherapy. Biopsy specimens taken both from stomach and colon were completely normal. Thereafter, she received a total of six cycles of CHOP in the outpatient clinic. Twelve months after discharge, the patient remains well except a mild paresthesia on both lower limbs.

DISCUSSION

POEMS syndrome also known as Crow fucaise syndrome, was first described in Japan as a rare multisystem disease

Figure 1 A snakeskin-like appearance of gastric mucosa.
characterised by polyneuropathy, organomegaly, endocrinopathy, monoclonal gammapathy, and skin lesions. Our patient fulfilled all of the above mentioned characteristic criteria and the diagnosis of POEMS syndrome was made.

Gastrointestinal involvement is not a part of the syndrome but gastrointestinal symptoms may be an accompanying clinical feature. There are many cases of POEMS syndrome in the literature but none have suggested gastrointestinal involvement without amyloid. Our patient had snakeskin-like appearance in the upper endoscopy. This form of a gastric appearance is probably related to POEMS syndrome. Preliminary data suggest that raised vascular mediators. Similarly these mechanisms have also been implicated in the pathogenesis of colitis thus we strongly believe that the colitis of our patient could well have been attributable to the immune dysregulation and cytokine induced intestinal epithelial cell apoptosis.

The prognosis for patients with POEMS has been reported to be poor, with median survivals estimated to be 12 to 165 months. Survival is not affected by the number of POEMS features. Clubbing, extravascular volume overload, stroke, and myocardial infarction are associated with shorter survival. Because the pathogenesis of this multisystem disease is elusive, treatment is not standardised.

Overall we advocate that patients with POEMS syndrome who have gastrointestinal complaints should thoroughly be evaluated for gastrointestinal involvement. We also highlight the necessity of further studies for better delineation of the characteristics of this entity in POEMS syndrome.

Figure 2 Colonoscopic biopsy specimen that shows neutrophilic infiltration of the lamina propria consistent with colitis.