

中文題目：慢性腸道假阻塞發生於一位進行性全身性硬化症病人：一病例報告與文獻回顧

英文題目：**Chronic Intestinal Pseudo-obstruction in a Patient with Progressive Systemic Sclerosis — A Case Report and Review of the Literature**

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### **Introduction:**

Progressive systemic sclerosis (PSS) is a systemic disease of uncertain cause and is characterized by widespread collagen deposition that results in tissue fibrosis. Skin involvement can be limited to the face and distal extremities (limited type), or it can be diffuse to the trunk (diffuse type). Limited type PSS is frequently associated with CREST (ie, calcinosis, Raynaud phenomenon, esophageal disease, sclerodactyly, and telangiectasis) syndrome. Gastrointestinal involvement manifests in up to 90% of patients with PSS. The pathologic findings are smooth muscle atrophy and fibrosis caused by collagen deposition mainly in the tunica muscularis. Any part of the gut may be involved. Hypomotility from smooth muscle atrophy and fibrosis causes stasis, dilatation, and intestinal pseudo-obstruction. Here we report a patient with limited type PSS and chronic intestinal pseudo-obstruction (CIPO).

### **Case report:**

A 69-year-old woman presented with severe abdominal pain and distension for 2 days. She is a patient with limited type PSS. She had experienced the same episodes in the past few months. Physical examination demonstrated Raynaud phenomenon, sclerodactyly, skin tightness on face, hands and forearms, polygonal telangiectasis on face and hands, soft and distended abdomen with diffuse tenderness, tympany to percussion and a paucity of bowel sounds. There were no signs of mechanical intestinal obstruction or peritonitis. The radiographic finding of abdomen is severe dilated loops of small bowel, termed the hidebound bowel sign. She was treated conservatively with parenteral nutritional support, cisapride and antibiotics.

### **Discussion:**

Chronic intestinal pseudo-obstruction is a rare entity characterized by recurrent attacks of intestinal obstruction without mechanical causes. The diagnosis is based on history and radiographic findings. It is frequently associated with PSS. Smooth muscle atrophy and fibrosis result in clinical manifestations and the hidebound bowel sign. Conservational treatments, such as parenteral nutritional support, medications that enhance gastric motility, or inhibit bacterial overgrowth, are usually not satisfied. Effective treatment with subcutaneous octreotide for CIPO was reported in limited cases.