Case Report

Normal Bowel Function Restored After Oxygen Therapy in Systemic Sclerosis and Colonic Inertia

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ABSTRACT. Colorectal involvement with obstructed defecation is a common complication of progressive systemic sclerosis (pSSc), compromising quality of life and placing patients at risk for life-threatening complications. Treatment for colonic inertia in these patients includes laxatives, prokinetics, and ultimately colectomy, which is associated with high morbidity and mortality in pSSc. We describe a woman with scleroderma and colonic inertia recommended for total abdominal colectomy. As a result of respiratory decompensation, she was placed on oxygen by nasal cannula, after which bowel motility with regularity was restored, obviating the need for colectomy. (First Release July 1 2007; J Rheumatol 2007; 34:1777–8)

Key Indexing Terms:
PROGRESSIVE SYSTEMIC SCLEROSIS SCLERODERMA BOWEL FUNCTION
OXYGEN THERAPY CONSTIPATION COLONIC INERTIA COLECTOMY

Colorectal involvement with either fecal incontinence or obstructed defecation is a recognized feature of progressive systemic sclerosis (pSSc). It is often underestimated and compromises quality of life in as many as 20% of patients with scleroderma[1]. Constipation has a documented prevalence of ~30%, with overall necessity of digital evacuation being as high as 18% (also underreported)[1,2]. Complications include bleeding from abraded telangiectasia and stercoral ulcers, perforation, and infection. Slow transit constipation (STC) or colonic inertia is often treated with laxatives, prokinetic agents such as 5HT agonists and erythromycin, and, at the extreme, colectomy with ileorectal anastomosis (IRA). Total colectomy with IRA is a common operative procedure in the general population with STC who fail to respond to conservative treatment[3]. The procedure carries a high satisfaction rate by patient report despite concomitant complications such as small bowel obstruction (~20%), persistent constipation (6%–33%), diarrhea or frequency (~40%), fecal incontinence (13%–47%), anastomosis leakage, and need for repeat surgery[3-7]. Patients with scleroderma are more likely to experience postsurgical complications of colectomy, being poor surgical candidates due to underlying cardiovascular, pulmonary, and renal disease as well as inherent issues related to pSSc such as impaired healing, systemic steroid treatment, compromised bowel wall integrity, and potential vasculitis[8,9]. In 1962, the first successful partial colectomy for STC in scleroderma was reported[10] and by 1976 a total of only 5 successful cases had been reported[11], with most attempts in scleroderma patients ending in failure and death[8,11]. Although colectomy has been increasingly more successful in pSSc, outcomes are still poor in these patients, with cautious approach and segmental resection recommended if conditions permit[8].

CASE REPORT

A 53-year-old Caucasian woman first had manifestations of pSSc in 1996, with a subsequent 4-year history of intermittent episodes of dyspnea, skin thickening with mild sclerodactyly, Raynaud's syndrome, and constipation alleviated with aggressive daily laxative use. Diagnosis of scleroderma was made in 2000 after skin biopsy, with subsequent referral to Rheumatology. By 2003, constipation progressed to a severe intractable state, with bowel movements occurring every 12 days only with manual disimpaction of hard stool. Treatment included prokinetic agents and laxatives, without amelioration. Bloating with increased weight gain was also reported, but the patient never experienced dysphagia. Plaquenil 200 mg twice daily was the only medication. She ate small normal fiber meals and was compliant with fluid intake. Examination revealed normal rectal sphincter tone with absence of masses. Sitz marker study revealed delayed transit with 23 out of 24 radiopaque markers 5 days after oral ingestion, retained in the left colon with rectosigmoid predominance, as revealed by single anteroposterior abdominal radiograph obtained 5 days after ingestion of markers. A defogram showing the rectum as opacified and balloon expulsion tests revealed normal evacuation of rectum. Colonoscopy revealed a normal mucosa without telangiectasias. A diagnosis of colonic inertia was made in 2004, with total abdominal colectomy with ileorectal anastomosis being offered. Colorectal consultants believed that based on our patient's significant symptoms, the benefits of surgery would greatly outweigh the increased risk of surgical procedure.

In 2005 she developed severe respiratory symptoms due to scleroderma-related fibrosis and was admitted to hospital. On admission, she had PaO_2 of 80 mm Hg and she was placed on oxygen via nasal cannula for the first time. Within 3–4 days, she produced a bowel movement without aid of laxative, prokinetic agent, or manual assistance. At the time of hospital discharge, daily

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bowel movements were maintained with use of nocturnal oxygen at 2 l/min for 6 to 8 hours. After this sequence of events, she received cyclophosphamide for treatment of pulmonary fibrosis.

One year later, nocturnal oxygen use was stopped due to lack of pulmonary oxygen requirement. This resulted in progressively decreased frequency of bowel movements, returning to the point of previous severity over a 3-month period. Reinitiation of oxygen restored daily bowel movements within a few days. Prior to the initial use of oxygen and during periods without use of nocturnal oxygen, the patient described her bowels as "asleep, dead"; with oxygen she reports the ability to sense "waves" in her abdomen (peristalsis). In 2007, regularity is maintained with continued use of nocturnal oxygen. Total abdominal colectomy was not performed.

DISCUSSION

In pSSc, pathology of the gastrointestinal (GI) tract is similar throughout. Smooth muscle atrophies and is replaced with collagen deposition and fibrosis setting into the component layers of the GI tract. In the lower GI tract, this results in weakening of the bowel wall, often causing the wide-mouth antimesenteric sacculations seen on gross pathology and barium enema. The pathophysiology of pSSc-related GI manifestations is not well understood, but is characterized by the same neuronal and vascular dysfunction, inflammation, and fibrosis found in other organ systems affected by pSSc. Similar to the body of evidence related to pulmonary and digital ischemic compromise, Jaovisidha and colleagues summarize evidence pointing to mucosal hypoxia and vascular insufficiency as the potential lead insult to GI smooth muscle[12]. Such observations allow if conditions permit for an understanding of the potential role of oxygen therapy in pSSc-related GI dysmotility.

We invite correspondence relating similar clinical experiences. Further investigation into oxygen-responsive colonic dysmotility in pSSc should be undertaken. We encourage specialists to attempt a trial of oxygen, a benign, noninvasive, and low-cost intervention, in patients with pSSc-related STC in nonacute settings before considering extreme and potentially harmful options such as elective colorectal surgery.

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