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Crohn's disease of the larynx

GERARD J. GIANOLI, M.D., ROBERT H. MILLER, M.D.

Abstract
Extraintestinal involvement of Crohn's disease is becoming more readily identified. Laryngeal involvement by Crohn's disease, however, has been reported in only five cases in the medical literature. We present the sixth case as well as an analysis of the prior reports.

Key words: Crohn disease; Laryngeal diseases

Introduction

Crohn's disease, or regional enteritis, is characterized by granulomatous inflammatory lesions that typically involve the distal small bowel and variable segments of the colon. The disease process may, however, involve any portion of the gastrointestinal tract. The lesions tend to be focal with skipped areas of normal mucosa. Extraintestinal manifestations of regional enteritis were not appreciated by Crohn (Crohn *et al.*, 1932) in his early description of the disease in 1932, but since then many reports have identified extraintestinal sites involved with Crohn's disease. These include the joints, liver, anus, skin, bone marrow, eye, oesophagus, and oral cavity.

One of the most common head and neck manifestations of Crohn's disease is oral ulceration and oedema. However, other head and neck sites may become involved. Crohn's disease involving the larynx is very rare and only five cases have been reported in the medical literature (Croft and Wilkinson, 1972; Kelly *et al.*, 1979; Ramsdell *et al.*, 1984; Lemann *et al.*, 1987). We report the sixth case and have reviewed the previous reports for analysis.

Case report

The patient a 44-year-old black female, was in her usual state of health until 14 months prior to admission when she discontinued prednisone therapy for her longstanding Crohn's disease. shortly thereafter she began to experience painful genital ulcers. Approximately 13 months prior to admission painful oral ulcerations appeared and then problems with odynophagia and hoarseness began. Twelve months prior to admission the patient also noted increasing difficulty with bloody diarrhoea. Three to four months prior to admission she began to notice nasal crusting and bloody nasal discharge. Five days prior to admission the odynophagia had become so severe that oral intake was limited to selected liquids only. The day of admission the patient presented to the Charity Hospital of New Orleans' Delgado Sexually Transmitted Disease Clinic due to concern about the progressive genital and oral ulcers. The patient was admitted to the gynaecology service for evaluation and the otolaryngology–head and neck surgery service was consulted concerning the oral ulcerations and odynophagia.

Her past medical history was significant for well controlled schizophrenia, asthma, and Crohn's disease diagnosed seven years previously. She had undergone a hysterectomy and bilateral salpingoopherectomy, had had four uncomplicated spontaneous vaginal deliveries in the past and suffered with symptoms of stress urinary incontinence. Medications included haloperidol, sulphasalazine, ranitidine, diphenhydramine, oestrogen, theophylline, albuterol, and biperiden. There were no known allergies. The patient had smoked one to one and a half packs of cigarettes a day for the past 30 years and was sexually active.

On physical examination the patient was an obese black female in no distress with a very breathy voice. Her hair was thin with areas of alopecia. The oral cavity revealed a large ulcer on the hard palate and another over the left maxillary gingiva (Figure 1). The nasal examination using endoscopy revealed much crusting, friable mucosa, and a few ulcerated lesions. Laryngeal examination displayed normal vocal fold mobility but marked oedema of the posterior commissure and arytenoid area was present. Additionally, ulcerations were present on the right mid true vocal fold and opposing ulcers were present on the true folds posteriorly. The epiglottis was ulcerated and cobblestone in appearance. Ulcerative lesions were also present on the vulva and perianal regions. The remainder of the physical examination was normal.

Results of serum chemistries were normal, as well as the routine coagulation profile. Complete blood cell count revealed 13 100 white blood cells, 9.3 g haemoglobin, and 27 per cent haematocrit. The theophylline level was 0.9 μg/ml. Urinalysis and urine culture were negative. Serological tests for syphilis including an ART, TPHA and a dark-field examination were negative. Culture and isolation studies were negative for toxoplasmosis, herpes simplex, and chancroid. Stool examination was negative for ova, parasites, and clostridium difficile toxin. Barium contrast examinations of the gastrointestinal tract demonstrated marked cobblestoning of the transverse and ascending colon with complete loss of normal mucosal pattern, consistent with Crohn's disease.

Biopsy of the oral lesions revealed nonspecific inflammation. Biopsy of the vulvar lesions demonstrated chronic erosive vulvitis with fibrosis and lipoatrophy. A left colon biopsy displayed acute and chronic colitis with regional depletion of goblet cells.

Therapy consisted of antibiotics, local wound care, sitz baths, bladder catheterization, nutritional support, sulphasalazine and

From the Department of Otolaryngology–Head and Neck Surgery, Tulane University School of Medicine, New Orleans, Louisiana, USA.
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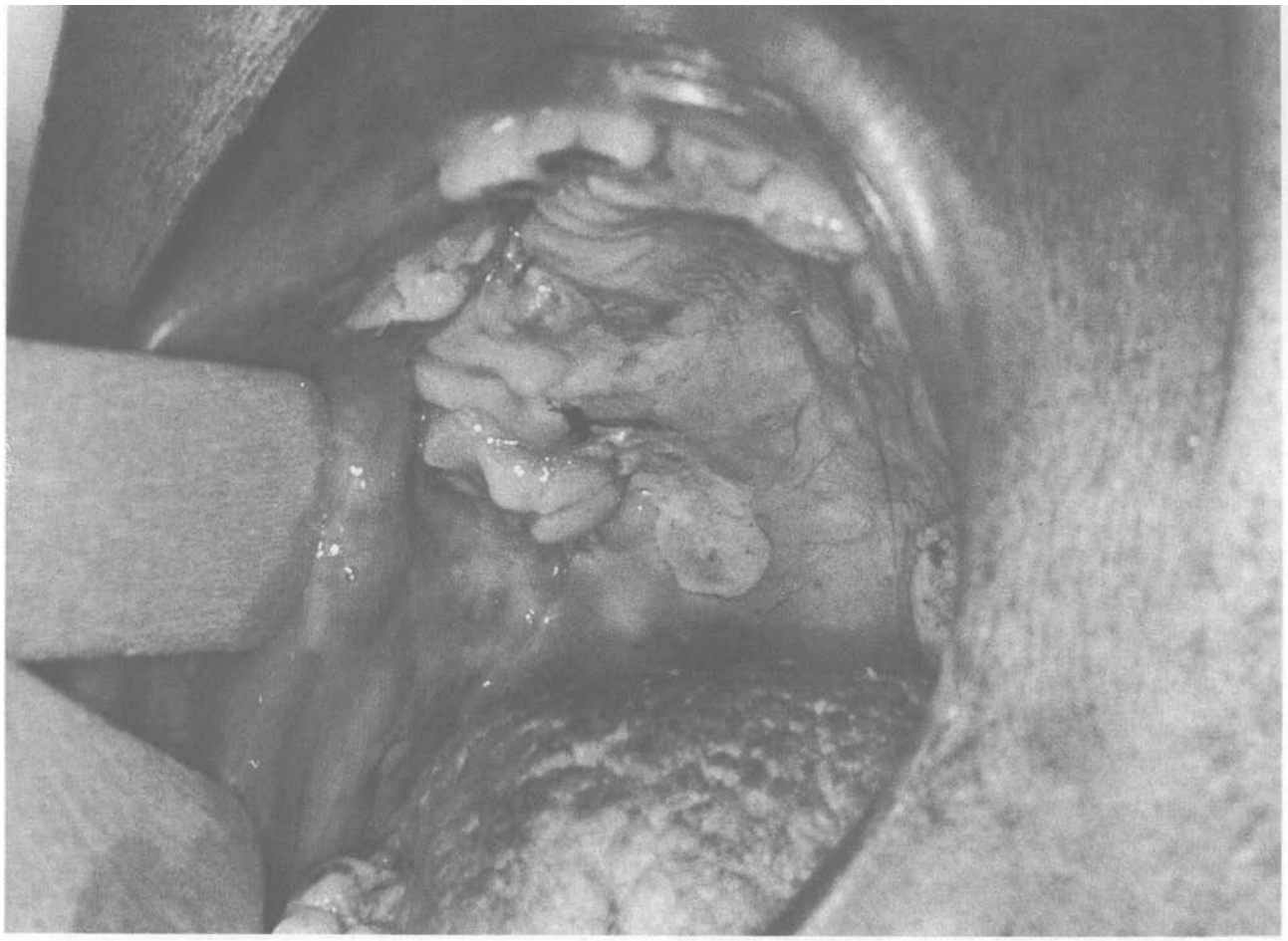

FIG. 1
Ulcerative oral lesions.

prednisone. Clinical improvement was noted just two days after starting prednisone therapy and there was marked improvement by the time of discharge (four days later). At the one month follow-up visit all ulcerative lesions and clinical symptoms had resolved. The patient has since been lost to follow-up.

Discussion

Crohn's disease is most typically manifested by abdominal pain, mild nonbloody diarrhoea, anorexia, and mild anaemia. The gastrointestinal symptoms are usually present for several years before the diagnosis is made due to the 'smouldering' chronicity of this disorder. Not uncommonly abdominal pain may precede the diarrhoea and it is invariably located in the lower abdomen. Generalized fatigue may be so severe so as to preclude normal daily activities. Anorexia and nutritional deficiencies may be marked in these patients. (Rosenberg, 1982).

Although initially thought to be a purely 'intestinal' disorder of the small bowel, many other systems have been noted to become involved. The skin, joints, liver, bone marrow, eye, anus, oesophagus, oral cavity, and almost any portion of the gastrointestinal tract have been affected by Crohn's disease.

Laryngeal involvement is a very unusual manifestation of Crohn's disease. Only five cases have been documented in the medical literature. Review of the reported cases (Tables I and II) allows some insight into detecting future cases and possibly preventing ill sequelae. Croft and Wilkinson (1972) were the first to describe Crohn's disease affecting the larynx. Laryngeal examination revealed generalized oedema, ulcerations, erythema, and granulation tissue with normal appearing false vocal folds. The laryngeal involvement was apparently brought to the attention of the physicians due to the marked dysphagia, odynophagia, and blood-tinged expectorate. Marked improvement in laryngeal symptoms was obtained with systemic steroid therapy.

Kelly *et al.* (1979) reported two cases of Crohn's disease of the larynx. These patients presented with symptoms of airway obstruction including stridor, dyspnoea, and nocturnal dyspnoea. Dysphagia, odynophagia, hoarseness, and otalgia were also presenting complaints. Although both patients improved with steriod therapy, one patient has required tracheotomy for

TABLE I

Author	Age (years)	Sex	Outcome	Larynx
Ramsdell *et al.* (1984)	12	F	Resolved	Ulcers
Kelly *et al.* (1979)	25	M	Resolved	Oedema, VC adduction, obstruction
Kelly *et al.* (1979)	22	F	Trach	Oedema, obstruction
Croft and Wilkinson (1972)	27	M	Improved	Oedema, ulcer, granulation tissue
Lemann *et al.* (1987)	24	F	Resolved	Oedema
Present study	44	F	Resolved	Oedema, red, ulcers

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TABLE II
SITES OF INVOLVEMENT

Author	Gastrointestinal	Skin	Joint	Genital	Oral	Bronchial	Nasal
Ramsdell *et al.* (1984)	+	+	+	+	+	0	0
Kelly *et al.* (1979)	+	0	0	0	0	0	0
Kelly *et al.* (1979)	+	0	+	0	+	0	+
Croft and Wilkinson (1972)	+	0	0	0	+	0	0
Lemann *et al.* (1987)	+	+	0	0	+	+	+
Present study	+	+	0	+	+	?	+

airway control. Airway obstruction was attributed to cricoarytenoid joint involvement and extensive oedema. Ramsdell *et al.* (1984) described a case of Crohn's disease with multiple system involvement that displayed laryngeal ulcerations. This patient had symptoms of hoarseness and dyspnoea which promptly resolved after steroid treatment.

Lemann *et al.* (1987) reported on two patients with bronchial involvement with Crohn's disease of which one affected the larynx. The patient experienced aphonia, dysphagia, and dyspnoea. The larynx was simply described as oedematous, although the bronchi displayed erythema and numerous whitish granulations. Again, the symptoms resolved rather dramatically with systemic steroid treatment.

All cases occurred in young patients, generally in their twenties. All patients had experienced the gastrointestinal symptoms of Crohn's disease prior to laryngeal symptoms and most had multiple extraintestinal manifestations of Crohn's disease. All but one patient had dramatic relief after appropriate steroid treatment.

Treatment of laryngeal Crohn's disease may involve emergency airway intervention such as endotracheal intubation or tracheotomy. Oxygen administration via a cool mist vapourizer is recommended. Immediate administration of corticosteroids in pharmacological doses should be performed whenever airway compromise is suspected. Appropriate consultation with the gastroenterology service may be invaluable in the patient with undiagnosed Crohn's disease. Prognosis for laryngeal involvement appears to be very good with timely administration of treatment.

Conclusions

Although Crohn's disease of the larynx is rare, it is an important entity to identify due to possible severe sequelae. The patient with laryngeal involvement of Crohn's disease will typically have previously diagnosed Crohn's disease. Odynophagia, dysphagia, and hoarseness will be the primary complaints. However, airway distress may be present in varying degrees. Laryngeal examination will reveal erythema, oedema, and ulceration. Oral cavity involvement with ulceration and oedema is present in most cases and is typically the most significant physical finding on initial examination. Other extraintestinal manifestations may be present and steroid therapy should bring about prompt resolution of all lesions.

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Address for correspondence:
Gerard J. Gianoli, M.D.,
Department of Otolaryngology–Head and Neck Surgery,
Tulane University School of Medicine,
New Orleans, Louisiana 70112,
USA.

Fax: (504) 582–7846