Oral Crohn's disease : a case report with review of the literature

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ABSTRACT :

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Oral manifestation can be the first and an exclusive presenting symptom of Crohn's disease, a chronic inflammatory granulomatous disease which is uncommon in Thailand. Many types of lesions were described but the most frequent oral finding is nonspecific recurrent aphthous ulcer. We herein report a case of 30-year-old Thai female with oral Crohn's disease who presented with intractable recurrent painful oral ulcers for many years preceding non-specific gastrointestinal tract symptoms.

Key words: Crohn's disease, Oral manifestations, Oral ulcers

บทคัดย่อ :

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สาขาวิชาตจวิทยา ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย

รอยโรคในช่องปากอาจเป็นอาการแสดงแรกและอาการเดียวที่พบในโรคโครนส์ (Crohn's disease) ซึ่งเป็นโรคลำไส้อักเสบชนิดหนึ่งที่พบไม่บ่อยในประชากรไทย โรคนี้มีการตรวจพบการอักเสบตลอดผนังลำไส้เป็นชนิดแกรนูโลมาและมีอาการที่พบในปากหลายชนิด ชนิดที่พบมากที่สุดเป็นแผลร้อนใน (aphthous ulcers) ที่กลับเป็นซ้ำบ่อยๆ รายงานฉบับนี้นำเสนอผู้ป่วยหญิงอายุ 30 ปี ที่มาพบแพทย์ด้วยแผลเจ็บในเยื่อบุช่องปากเป็นๆ หายๆ ตามมาด้วยอาการที่ไม่จำเพาะของระบบทางเดินอาหาร

คำสำคัญ: โรค Crohn's, อาการแสดงทางปาก, แผลในปาก

Introduction

Crohn's disease (CD) is characterized by chronic, transmural granulomatous inflammation that may involve any segment of the gastrointestinal tract from the mouth to anus. Oral manifestations are divided into 2 groups. The first one is non-specific lesions which are more common and the second one being specific lesions which demonstrate macroscopic and microscopic changes similar to that of the gastrointestinal tract.

Case report

A 30-year-old woman presented with painful recurrent oral ulcers for 3 years with no other mucocutaneous lesions. Her lesions were mostly self-limited, otherwise healed promptly after a short course of oral prednisolone. Her medical history was unremarkable except for occasionally having watery diarrhea which spontaneously resolved. No history of passing mucous bloody stool, fever or weight loss. Oral examination revealed a large ulcer with irregular border, surrounded by erythematous halo. Its base was covered with yellowish pseudomembrane and also a whitish scar was observed on the hard palate (Figure 1 a-b). No genital ulcer was detected.

Laboratory examinations including complete blood count, urinary analysis and screening antinuclear antibodies (ANA) were within normal limits. Histopathological examination from oral ulcer showed fibrous stroma that was infiltrated by numerous lymphocytes, eosinophils, plasma cells and epithelioid histiocytes with necrotic tissue. The additional staining with GMS and AFB were negative (Figure 2 a-b). No immune deposition was detected in the skin specimen using direct immunofluorescence technique.

Figure 1a

Figure 1b

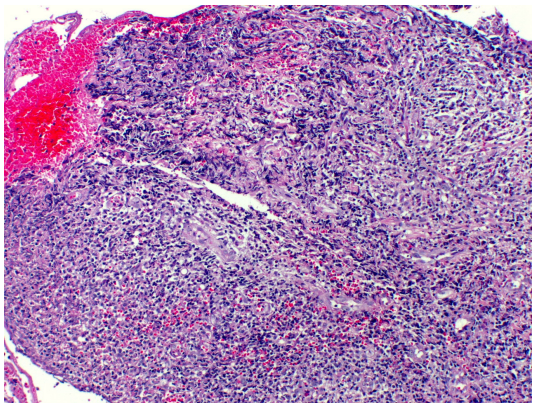

Figure 2a

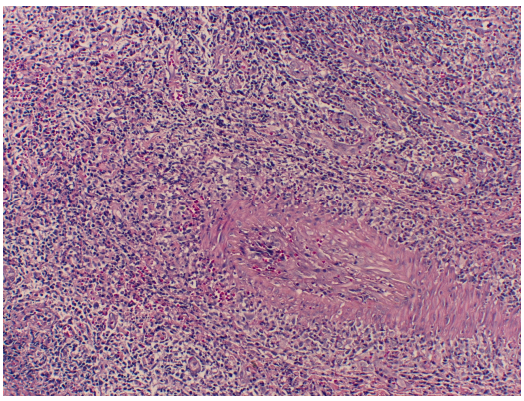

Figure 2b

The patient was sent for esophagogastroduodenoscopy and colonoscopy which revealed 2 large clean-based ulcers at ileocecal valve size 0.8 and 1.0 cm respectively. The histopathology showed chronic active colitis with ulcer. After ruling out infectious causes, the diagnosis of Crohn's disease (CD) was made. The patient was treated with oral prednisolone 40 mg/day and azathioprine 50 mg/day with complete response.

Discussion:

The prevalence of oral lesions in patients with CD vary from 0.5-80 %. They may be the presenting symptom and sign in 5-60% of patients.[1-6] All age groups can be affected but they are more common in children.[3] Sometimes, it is the sole manifestation.[3] A-16-year-review study of Thai patients with inflammatory bowel disease reveals only a single CD case with oral ulcer, out of a total of 10 patients with CD.[7]

Many oral lesions described in inflammatory bowel diseases including CD and ulcerative colitis (UC) are non-specific, such as aphthous ulcer, angular cheilitis, facial edema, gingivitis or gingival erythema. While macrocheilia with or without fissuring, cobblestoning of oral mucosa, deep linear ulcers of buccal vestibules, polypoid mucosal tags and pyostomatitis vegetans are regarded as specific and pathognomonic.[2,3] Oral lesions of CD was first described by Dudeney in 1969.[8] The specific oral ulcers of CD are characterized by deep, linear ulcers with hyperplastic folds that locate in oral vestibules or gingival sulci.[1,4] On the other hand, Pyostomatitis vegetans is more common in UC[1] and characterized by multiple friable pustules, erosions and ulcerations on erythematous background with vegetations on the gingiva or buccal mucosa.

Histopathologic findings in oral CD show high incidence of non-caseating granuloma (75-100%), despite their various presentations.[5,9,10] Special stains are used to rule out other causes of granulomatous inflammation such as deep fungal infections, tuberculosis, and foreign body reactions.[9,10]

Our patient presents with chronic painful irregular large oral ulcers. Differential diagnosis should include chronic ulcerative stomatitis, eosinophilic ulcer, major aphthous ulcer, infectious causes, systemic causes (i.e. inflammatory bowel disease) and squamous cell carcinoma. A biopsy taken from the lesion reveals non-caseating granuloma without evidence of organism on special stains. In addition, she has a history of having

intermittent watery diarrhea in a few years. Colonoscopy demonstrates colon ulcers at ileocecal valve. Tuberculosis is excluded by histopathology. The diagnosis of CD is then entertained.

Even though some studies suggest that the upper gastrointestinal tract involvements are more common in patients with oral CD,[4,11] The prospective study do not show a definite association between patients with oral CD and type of gastrointestinal disease.[5] In addition, oral lesions can persist or resolve regardless of the disease activity.[12]

Some of the lesions may spontaneously resolve, as seen in the patient. For mild-to-moderate symptomatic cases, topical beclomethasone mouth washes and 5-aminosalicylic ointments or sprays are also effective. For deep painful ulcers, slow-release intralesional corticosteroids are suggested. In refractory case, topical tacrolimus and systemic corticosteroids are recommended for short term usage. Choices for long term treatments are biologic therapies including infliximab, thalidomide with or without methotrexate and clofazimine.[10] We treats our patient with systemic steroids and azathioprine and her ulcers rapidly regress.

In conclusion, we encounter a young Thai patient with oral Crohn's disease in which diarrhea is a clue to diagnosis. Crohn's disease is not common in Thailand and oral involvement may be the only presentation. Thus, a high index of suspicion for the condition in patients with intractable oral lesions is very important.

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