Pyoderma gangrenosum in a patient with primary sclerosing cholangitis: A rare association

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Abstract

Pyoderma gangrenosum is a rare clinical entity with cutaneous ulcerations and is associated with systemic diseases. Inflammatory bowel disease is a common association. Pyoderma gangrenosum associated with inflammatory bowel disease and primary sclerosing cholangitis (PSC) has been reported. But Pyoderma gangrenosum associated only with PSC is not reported. We report a case of 57 year old lady with Pyoderma gangrenosum associated with PSC alone.

Introduction

Pyoderma gangrenosum is a rare type of cutaneous ulcers. It is a rapidly spreading painful ulcer with violaceous overhanging and undermined edges with necrotic or purulent base sometimes exposing underlying muscles or tendons. PG was first described by a French dermatologist Brocq who considered it as a bacterial infection. Even though its pathogenesis is still unkown, it is now categorized as a neutrophilic dermatosus with significant immune defect in cell-mediated immunity of neutrophil and monocyte function and humoral immunity. Associated systemic disease is seen in 50% of cases. Common associations are inflammatory bowel disease, sero-positive and sero-negative arthritis and monoclonal gammopathies. It is also associated with autoimmune diseases like lupus erythematosus and Sjögren syndrome[8]. Remaining cases are idiopathic. Although one case has been reported as a case of PG associated with both ulcerative colitis and PSC[2], we could not find a case of pyoderma gangrenosum associated with PSC alone. Therefore this may be the first case of pyoderma gangrenosum associated with primary sclerosing cholangitis alone.

Case report

A 57 year old lady from Batticaloa, in the eastern province of Sri Lanka presented with alteration of bowel habits, epigastric burning pain and jaundice over three months duration. Further evaluation of the history and clinical examination suggested an obtructive jaundice. Serum biochemical profile confirmed the clinical diagnosis.Ultrasound scan and contrast enhanced Computer Tomography suggested primary sclerosing cholangitis as the cause for obstructive jaundice. Endoscopic Retrograde Cholangio Pancreatography was performed at gatro-enterology unit and confirmed the diagnosis of PSC. Upper and lower gastro intestinal endoscopic findings were not showing any changes suggestive of inflammatory bowel disease. Multiple biopsies from gastro intestinal tract further excluded inflammatory bowel disease. She developed multiple bullous lesions (Figure 1) over anterior aspect of right shin, right knee and over dorsal aspect of the right foot while she was on treatment for PSC. Bullae ruptured resulting in painful ulcers. On examination the ulcers were having violaceous undermined edges with necrotic and granulating base (Figure 2). Clinical diagnosis of vesiocobullous type of PG was supported by the inflammatory changes on histology. Biochemical investigations are shown below.

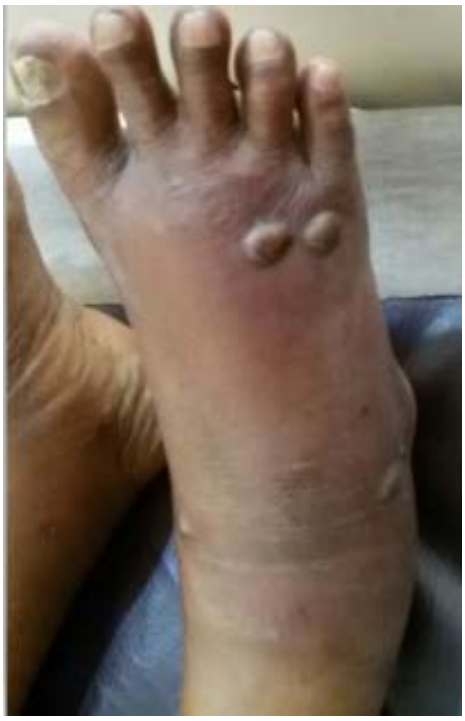

Figure 1. *Bullous lesions.*

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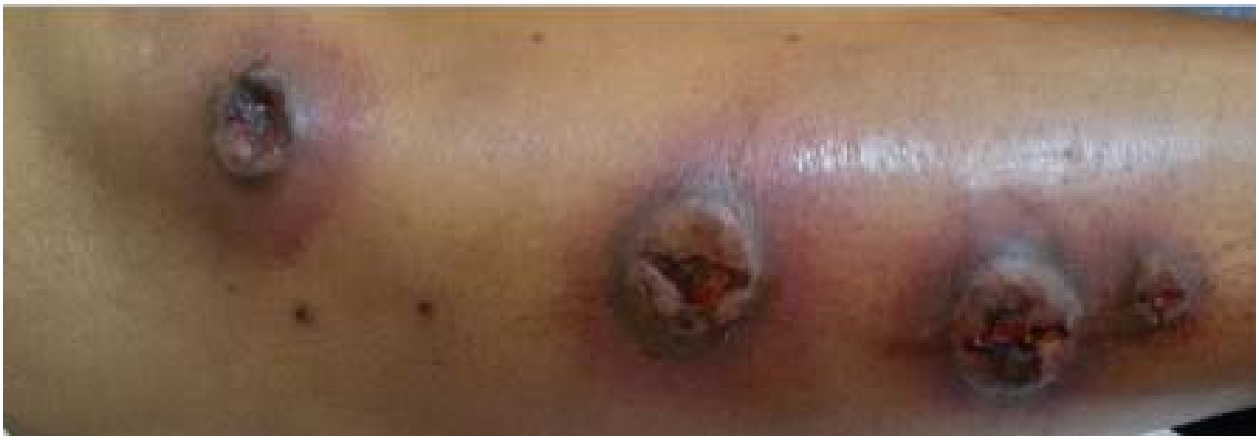

Figure 2. *Ulceration of bullous lesions.*

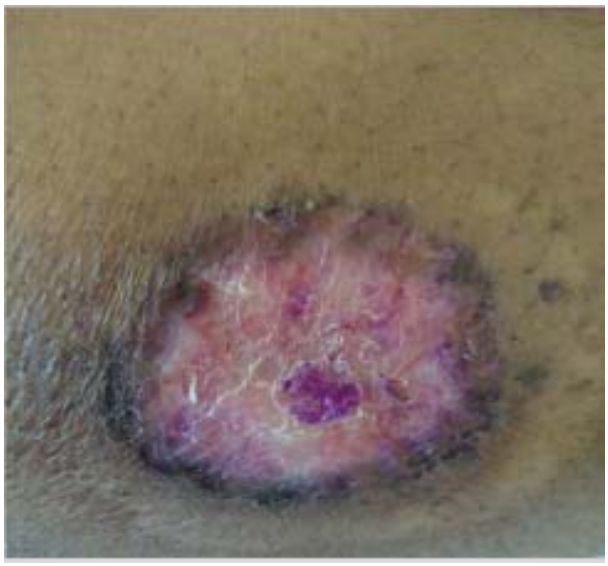

Figure 2. A resolving ulcer.

Blood and biochemical Investigations:

Hb 11.5 g/dl, Platelets 154x10^3, SGOT 160 iu/l, SGPT 85 iu/l, ALP 1400 u/l

Total Protein 7.7g/dl, Albumin 1.7g/dl, Globulin 5.7g/dl, Albumin/Globulin = 0.3

Total Bilirubin 219 umol/l, Direct Bilirubin 156 umol/l, S.creatinine 0.7mg/dl

Serum Na 136 meq/l, Serum K 3.8 meq/l

Her leg ulcers completely healed with the treatment of oral corticosteroids, which also supported the diagnosis of PG.

Discussion

Pyoderma gangrenosum is an inflammatory ulcerative condition of the skin, which is associated with systemic diseases. There is a slight female predominance. It usually presents in fourth or fifth decade of life. Its pathogenesis is still unkown. But it is now categorized as a neutrophilic dermatosus. There is underlying immune defect, seen in 25-50% of patients, especially in cell-mediated immunity, neutrophil and monocyte function. It also shows defects in humoral immunity.

There may be single or multiple ulcers, most commonly over pretibial area. But it can occur anywhere of the body. Unusual sites are vulva, penis, breast [1] and scrotum. Ulcer is the classical feature in PG. There are four clinical variants including pustular, vesicobullous, superficial granulomatous and pyostomatitis vegetans type. Bullous or haemorrhagic type of ulcerations are seen with drug intake or with hematological diseases. Acute myelogenous leukeamia or myelodysplasia and myeloproliferative disorders are associated with vesicobullous type. Chronic slowly enlarging ulcers with pustulation are

seen associated with inflamatory bowel diseases and athritis. Oral mucosal ulcerations have been reported with inflamatory bowel diseases. Ulcers may recur and residual cribriform type scarring is common. But prognosis of the disease is generally good if it is idiopathic or associated with treatable systemic disease. A significant number of patients follow a refractory course.

Pyoderma gangrenosum is commonly associated with inflammatory bowel diseases and 75% of primary sclerosing cholangitis patients show association with inflammatory bowel disease. Since our patient is 57 years old, the chances of diagnosing inflamatory bowel disease in her life is low. Therefore this patient is most probably having primary sclerosing cholangitis without inflammatory bowel disease giving rise to ulcers of pyoderma gangrenosum.

References

1. Spyridon Marinopoulos, Charalampos Theofanakis, Theoni Zacharouli, Maria Sotiropoulou, Constantine Dimitrakakis, Pyoderma Gangrenosum of the breast: A case report study. *Journal of the European Academy of Dermatology and Venereology* 2001; **12**: 257.
2. Goreti Catorze M, Pereira F, Fonseca F, Morbey A, Assis Pacheco F. Pyodermagangrenosum associated with sclerosingcholangitis, type 1 diabetes mellitus and ulcerative colitis. *Journal of the European Academy of Dermatology and Venereology* 2001; **15**: 257-9.
3. Sonya Martin Schwaegerle, Wilma F. Bergfeld, David Senitzer, Robert T. Tidrick. Pyodermagangrenosum. A review. *Journal of American Academy of Dermatology* 1988; **18**: 559-68.
4. Leon G, Clark BA, Stanislav N. Tolkachjov, Alina G. Bridges DO, Michael J. Camilleri MD. Pyosto matitisvegetans (PSV)-pyodermatitisvegetans (PDV). *A Clinico Pathologic Study* 2016; **75**: 578-84.
5. Rebecca Saich, Roger Chapman. Primary sclerosing cholangitis, autoimmune hepatitis and overlap syndromes in inflammatory bowel disease. *World Journal of Gastroenterology* 2008; **14**: 331-7.
6. Jisun Yoon, SeakHee Oh, Hyun Jin Kim, Sang Hyoung Park, Byong Duk Ye, Suk-Kyun Yang, Kyung Mo Kim. Primary Sclerosing Cholangitis with Inflammatory Bowel Disease in Korean Children. *Journal of Pediatric Gastroentero Hepato Nutrition* 2015; **18**: 268-75.
7. The odoros Androutsakos, Paraskevas Stamopoulos, Kiriaki Aroni, Gregorios Hatzis. A case report of successful treatment of pyodermagangrenosum in a patient with autoimmune hepatitis, and review of the literature. *BMC Gastroenterology* 2015; **15**: 149.
8. Sinéad M. Langan, Richard W Groves, Tim RCard, Martin C Gulliford. Incidence, Mortality, and Disease Associations of Pyoderma Gangrenosum in the United Kingdom: A Retrospective Cohort Study. *Journal of Investigative Dermatology* 2012; **132**: 2166-70.
9. RuiHua Shi, Bo Hao, ShunFuXu, QiYun Tang, JianXia Jiang. A patient with primary biliary cirrhosis and ulcerativecolitis with progression to primary sclerosing cholangitisand colorectal cancer: A case report. *International Journal of Case Reports and Images* 2012; **12**: 11-15.