A case report of Degos disease in patient presented with jejunal perforation and peritonitis.

Sujirod Hanthavichai MD,

Ratchathorn Panchaprateep MD,

Noppadon Noppakun MD.

ABSTRACT:

HANTHAVICHAI S, PANCHAPRATEEP R, NOPPAKUN N. A CASE REPORT OF DEGOS DISEASE IN PATIENT PRESENTED WITH JEJUNAL PERFORATION AND PERITONITIS. THAI J DERMATOL 2011; 27: 198-203.

DIVISION OF DERMATOLOGY, DEPARMENT OF MEDICINE, FACULTY OF MEDICINE, CHULALONGKORN UNIVERSITY, BANGKOK, THAILAND.

Degos disease or malignant atrophic papulosis is a progressive small- and medium-size arterial occluding disease, leading to tissue infarction in many organs such as the skin, gastrointestinal and nervous systems. This study reported a 56-year-old female patient who presented with abdominal pain due to jejunal perforation with peritonitis. Later, she developed generalized multiple central white scar and telangiectatic rim on trunk, extremities, palm and sole. The histopathology is characteristic, with wedge-shaped area of homogenized collagen fibers and lymphocytic vasculitis in the dermis. Unfortunately, she died from sepsis and multiorgan failure.

Key words: Degos disease, intestinal perforation

บทคัดย่อ :

สุจิโรตถ์ หาญทวิชัย, รัชต์ธร ปัญจประทีป, นภดล นพคุณ รายงานผู้ป่วยโรค DEGOS ที่มีอาการนำจากภาวะลำไส้ทะลุ และเยื่อบุช่องท้องอักเสบ วารสารโรคผิวหนัง 2554; 27: 198-203.

หน่วยโรคผิวหนัง ภาควิชาอายุรศาสตร์คณะแพทยศาสตร์จุฬาลงกรณ์มหาวิทยาลัย

โรค Degos เกิดจากความผิดปกติของหลอดเลือดแดงขนาดเล็ก และกลาง ทำให้มีการอุดตันของหลอดเลือดแดง และความผิดปกติในอวัยวะระบบต่างๆ เช่น ผิวหนัง, ทางเดินอาหาร และ ระบบประสาทส่วนกลาง เป็นต้น โดยรายงานฉบับนี้เป็นการนำเสนอ ผู้ป่วยหญิง อายุ 56 ปี ที่มาด้วยอาการปวดท้องจากภาวะลำไส้ทะลุ และเยื่อบุช่องท้องอักเสบภายหลังการผ่าตัด ได้สังเกตพบผื่นกระจายอยู่บริเวณ ลำตัว แขนขา ฝ่ามือและฝ่าเท้า โดย ตรงกลางรอยโรคมีแผลเป็น สีขาวจัดคล้ายสีขาวของเครื่องเคลือบหรืองาช้าง ล้อมรอบด้วย telangiectasia ที่บริเวณขอบของผื่น ร่วมกับลักษณะทางจุลพยาธิวิทยาแสดง wedge-shaped area of homogenized collagen และ lymphocytic vasculitis ในชั้นหนังแท้ ซึ่งเป็นลักษณะเฉพาะของโรค ภายหลังผู้ป่วยได้เสียชีวิตในเวลาต่อมา จากการติดเชื้อในกระแสโลหิต และอวัยวะภายในร่างกายล้มเหลว

คำสำคัญ: โรค Degos, ภาวะลำไส้ทะลุ

Introduction

Degos disease (DD) or malignant atrophic papulosis (MAP) was first described as a case of a disease by Kohlmeier in 1941. Then Degos was recognized as a distinct clinical entity in 1942. Degos disease is a rare disease with about 200 cases reported. All ages and races can be affected. The male to female ratio is approximately 3: 1. DD has a purely cutaneous variant and a systemic multiorgan variant[1]. Both have similar cutaneous eruptions. The natural history of DD has yet to be defined.

In this report, we present a patient who had a clinically fatal systemic Degos disease with typical cutaneous and characteristic histological manifestations.

Case report

The patient was a 56-year-old female from Nakornsawan presented with abdominal pain due to jejunal perforation with peritonitis. She underwent bowel resection surgery and anastomosis. After surgery, she developed sepsis and alteration of consciousness. Later she was referred to Chulalongkorn Hospital.

After admission in intensive care unit, attending physicans noticed that she had generalized multiple erythematous papules with central white scar on trunk, extremities, palm and sole. No history of other underlying disease was noted. None of the family members or close relative was affected with similar disorder or other dermatologic disease.

Physical examination revealed generalized multiple erythematous papules with central porcelain-white atrophic scar and crust with telangiectasia at rim on trunk, extremities, palms and sole (Figure 1.1-1.4). The lesions were blaching on pressure. She was drowsy and on endotracheal tube with respirator. Later, she was worsen and finally death from multiorgan failure and sepsis.

Figure 1.1

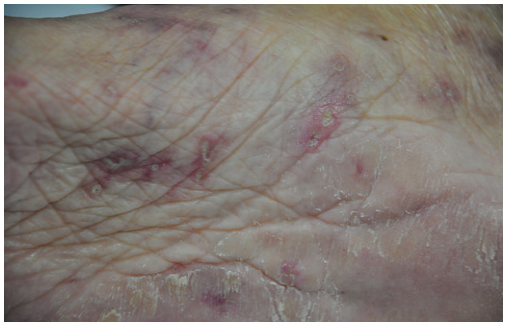

Figure 1.2

Figure 1.3

Figure 1.4

Figure 1.1-1.4 : Papules on trunk, extremities, palms and sole with central porcelain white atrophy and surrounding erythema.

Histopathological examinations from the right forearm and abdomen showed vacuolar degeneration of the basal cells, wedge-shaped area of homogenized collagen fibers in the dermis and perivascular lymphocytic infiltrate with intravascular platelet-fibrin thrombi (lymphocytic vasculitis). The Alcian blue stain is negative for mucin (Figure 2.1-2.3).

Laboratory findings: complete blood count (CBC) showed anemia and thrombocytopenia. Her coagulogram showed prolonged PT, PTT and INR.

Immunology profiles included ANA and anti-dsDNA was negative.

She was finally given a diagnosis of Degos disease or malignant atrophic papulosis.

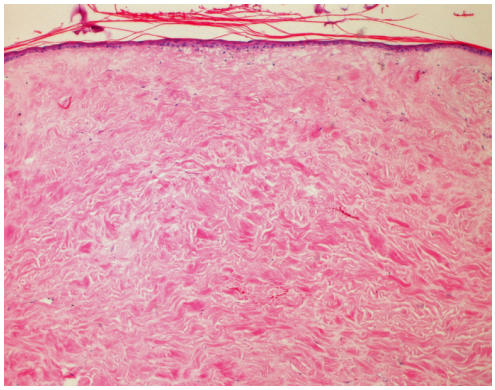

Figure 2.1 H&E sections show homogenized collagen fibers in the dermis.

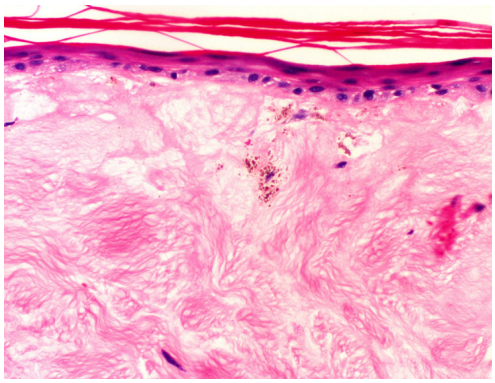

Figure 2.2 H&E sections show epidermal atrophy and vacuolar degeneration of the basal layer

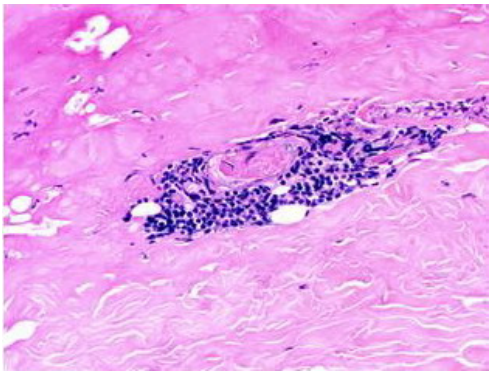

Figure 2.3 H&E sections reveal perivascular lymphocytic infiltrate with intravascular platelet-fibrin thrombi.

Autopsy was done and reported as acute necrotizing inflammation with multiple abscesses nearly entire small and large bowel from candidiasis, organizing peritonitis, multiple lung abscess at left upper lobe due to aspergillus infection, liver abscess at right lobe of liver, diffuse alveolar damage both lungs and acute tubular necrosis of both kidneys.

Discussion

Degos disease (DD) is an extremely rare and poorly understood disease. DD is an occlusive arteriopathy involving small-caliber vessels. It is a progressive, small- and medium-size arterial occluding disease, leading to tissue infarction. The possible mechanisms suggest disturbance in immunity, viral infection and abnormality in the clotting system of blood[1].

The cutaneous lesions of DD initially manifests with erythematous, pink or red papules, 0.5 – 1 cm in diameter. These papules heal to leave with pathognomonic central porcelain-white atrophic scar

and peripheral telangiectatic rim. The papules predominantly occur on the trunk and arms, rarely occurs on the face, palms and soles[2]. The lesions develop slowly and are usually asymptomatic, but may be accompanied by a slight burning sensation.

Systemic Degos disease can affect the gastrointestinal, nervous, opthalmological, cardiopulmonary and hepatorenal systems. Systemic Degos disease is frequently fatal, the patient usually die within 2-3 years from the onset of systemic involvement. Intestinal perforation is the most common cause of death. Other causes of death include bowel infarction, cardiopulmonary collapse, and neurologic infarction and hemorrhage. Systemic manifestations usually develop from weeks to years after the onset of skin lesions, or, in rare instances, they may precede the skin lesions[3,4].

No specific laboratory test can be used to aid in diagnosis of DD. In fact, most test results are normal, with the exception of anemia secondary to intestinal bleeding. Although, in some cases, antiphospholipid antibodies of uncertain significance are identified[5,6].

The characteristic histological findings demonstrate interface dermatitis, melanin incontinence and epidermal atrophy overlying a wedge-shaped area of cutaneous ischemia extending into the deep dermis. Marked endothelial swelling and occasional platelet-fibrin thrombi are often noted. Abundant acid mucopolysaccharides can be present in the dermis.

Differential diagnosis of the cutaneous manifestation include secondary vasculitis, Wegener granulomatosis, acute lupus erythematosus, connective tissue disease[7], atrophie blanche lesions and dermal mucinosis. Comprehensive blood analysis and examination of tissue pathology specimens is necessary to exclude diseases that mimic DD[8].

No medical therapy for DD is uniformly effective in previous reports. Antiplatelet drugs (eg, aspirin, dipyridamole) may reduce the number of new lesions in some patients with only skin involvement. Recent anecdotal studies reported roles of intravenous immunoglobulin[9] and eculizumab[10].

There is no specific treatment for systemic DD. This patient had severe sepsis, coagulopathy and thrombocytopenia and died from DIC with multiorgan failures due to multiple invasive fungal infections.

References

1. Scheinfeld N. Pairing and comparing nine diseases with Degos Disease (Malignant Atrophic Papulosis): an attempt to illustrate our understanding and direct future inquiry. Dermatol Online J 2009; 15: 10.
2. Notash AY, Mazoochy H, Mirshams M, Nikoo A. Lethal systemic Degos disease with prominent cardio-pulmonary involvement. Saudi Med J 2008; 29: 133-7.
3. Tan WP, Chio MT, Ng SK. Generalized red papules with gastrointestinal complications. Diagnosis: malignant atrophic papulosis (Degos' disease). Clin Exp Dermatol 2007; 32: 615-6.
4. Lankisch MR, Johst P, Scolapio JS, Fleming CR. Acute abdominal pain as a leading symptom for Degos' disease (malignant atrophic papulosis). Am J Gastroenterol 1999; 94: 1098-9
5. Asherson RA, Cervera R. Antiphospholipid syndrome. J Invest Dermatol 1993; 100: 21S-27S.
6. Saglik E, Baykal C, Buyukbabani N, Inanc M. Malignant atrophic papulosis: endocardial involvement and positive anticardiolipin antibodies. J EurAcadDermatolVenereol 2006; 20: 602-3.
7. Ortiz A, Ceccato F, Albertengo A, Roverano S, Iribas J, Paira S. Degos cutaneous disease with features of

connective tissue disease. J Clin Rheumatol 2010;16:132-4.

8. Scheinfeld N. Malignant atrophic papulosis.Clin Exp Dermatol 2007;32:483-7
9. Zhu KJ, Zhou Q, Lin AH, Lu ZM, Cheng H. The use of intravenous immunoglobulin in cutaneous and recurrent perforating intestinal Degos disease (malignant atrophic papulosis). Br J Dermatol 2007;157:206-7.
10. Magro C, Shapiro L, Johnson B, Salmon J, Whelan P. Rheumatology Grand Rounds Hospital for Special Surgery (New York City) Degos Disease. Feb 10, 2010. In press.