A Case of Degos Disease Complicated By Constrictive Pericarditis In Remote Phase

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Case report

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Abstract

Background: Degos disease, also known as malignant atrophic papulosis, is characterized by cutaneous manifestations due to chronic thrombo-obliterative vasculopathy. There have been reports of rare late-onset Degos disease complicated by constrictive pericarditis (CP). We report a case of CP caused by Degos disease that developed 20 years after diagnosis.

Case presentation: A 62-year-old woman who has been taking aspirin for 20 years for Degos disease was hospitalized for worsening heart failure. The patient was diagnosed with CP and underwent pericardiectomy. Pathological findings suggested the involvement of Degos disease. The postoperative course was uneventful, and her heart failure and Degos disease did not worsen.

Conclusions: This report suggests that Degos disease can cause long-term CP. Aspirin effectively inhibited the progression of Degos disease, and surgical treatment is necessary when heart failure due to CP is refractory to treatment.

Background

Degos disease, also known as malignant atrophic papulosis, is rare. To date, approximately 200 cases have been reported. Degos disease is characterized by cutaneous signs, such as central porcelain-white atrophic papules with an erythematous telangiectatic rim caused by chronic, thrombo-obliterative vasculopathy [1, 2]. There have been few reports of constrictive pericarditis (CP) caused by Degos disease [3, 4]. We performed a surgical intervention for CP caused by Degos disease, presenting with treatment-refractory heart failure. To the best of our knowledge, this is the first report of CP caused by Degos disease that developed 20 years after diagnosis.

Case Presentation

A 67-year-old woman was admitted to our cardiology department for dyspnea. Her medical history included hypertension, atrial fibrillation, diabetes mellitus, and Degos disease, and she had been taking low-dose aspirin for 20 years.

At the time of diagnosis, she exhibited cutaneous signs, and a histopathological examination displayed perivascular lymphocytic infiltration with distinct mucin deposition. These lesions were associated with Degos disease [1, 2]. No systemic symptoms were observed. Three years ago, gastrointestinal endoscopy revealed a small intestinal lesion, which was suspected as a systemic manifestation of Degos disease [5].

On admission, her blood pressure was 110/62 mmHg, and her heart rate was 99 beats/min with atrial fibrillation. Physical examination revealed liver enlargement, jugular vein distension with Kussmaul’s sign, and limb edema. Chest radiography revealed bilateral pleural effusion and calcification of the
pericardium. Computed tomography revealed bilateral pleural effusion and pericardial effusion with marked calcification of the pericardium (Fig. 1).

Cardiac catheterization revealed equal right and left ventricular end-diastolic pressures and square root signs (Fig. 2). No coronary artery stenosis was observed. Echocardiography revealed pericardial thickening, pericardial effusion, ventricular septal paradoxical motion, septal bounce, and a normal left ventricular ejection fraction. The cutaneous signs were similar to those observed 20 years ago. Endoscopy revealed the same findings 3 years previously [5].

Despite optimal medical treatment, her heart failure did not improve, and the patient became catecholamine-dependent. Therefore, surgical pericardiectomy was performed.

During the operation, the pericardium was markedly thickened and calcified. The pericardium was incised, and 200 ml of bloody fluid was suctioned. Inside the pericardial sac, there were adhesions with some calcification (Fig. 3A), partly infiltrating the myocardium (Fig. 3B). The thickened pericardium was then thoroughly resected.

The central venous pressure decreased from 30 to 16 mm Hg, and the cardiac diastolic capacity improved.

Histopathological examination of the pericardium revealed a high degree of fibrosis, vitrification, and calcification of the pericardium. Lymphocytic infiltration was observed around the pericardial vessels (Fig. 4A, B).

The postoperative course was uneventful. The patient was extubated on day 1, discharged from the intensive care unit on day 2, and discharged on day 18. After surgery, the patient received aspirin, furosemide, spironolactone, bisoprolol, and perindopril erbumine treatment for 4 years. Her heart failure has not worsened.

**Discussion And Conclusions**

Pierce et al. reported a case of chronic pleuritis and pericarditis in a 32-year-old woman with Degos disease [3]. In that report, the patient developed heart failure and required surgical treatment. Histopathological examination revealed a calcified and fibrotic epicardium, similar to our case, but there was no proliferative vasculitis of Degos disease. In our case, lymphocytic infiltration was present around the pericardial vessels, indicating the involvement of Degos disease.

According to Theodoridis et al., systemic signs were present in 29% of patients with Degos disease. Organ involvement began within the first 7 years of disease, and the mean survival time from the development of systemic disease was 0.9 years [2]. However, our patient, who took aspirin, did not develop a systemic disease until 17 years after diagnosis. Since the current study was a case report, a general conclusion cannot be made. However, Yukiiri et al. has reported CP caused by untreated Degos disease, medically treated with aspirin, dipyridamole, and furosemide [4].
Therefore, aspirin was found to effectively inhibit the progression of Degos disease. In summary, Degos disease can cause long-term CP. Aspirin effectively inhibits the progression of Degos disease, and surgical treatment is necessary when heart failure due to CP is refractory to treatment.

**Abbreviations**

CP, constrictive pericarditis

**Declarations**

**Ethics approval and consent to participate:** Ethics approval was not required for the retrospective analysis of this clinical case.

**Consent for publication:** Written consent was obtained from the patient.

**Availability of data and materials:** Not applicable.

**Competing interests:** The authors declare that they have no competing interests.

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**Authors’ contributions:** YK wrote the original draft. TK was in charge of writing, reviewing, and editing the manuscript. KM supervised the study. All authors read and approved the final manuscript.

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**References**

[1] Theodoridis A, Makrantonaki E, Zouboulis CC. Malignant atrophic papulosis (Köhleimer–Degos disease) – a review. Orphanet J Rare Dis. 2013;8:10.

[2] Theodoridis A, Konstantinidou A, Makrantonaki E, Zouboulis CC. Malignant and benign forms of atrophic papulosis (Köhleimer-Degos disease): systemic involvement determines the prognosis. Br J Dermatol. 2014;170:110-5.

[3] Pierce RN, Smith GJ. Intrathoracic manifestations of Degos’ disease (malignant atrophic papulosis). Chest. 1978;73:79-84.

[4] Yukiiri K, Mizushige K, Ueda T, Tomohiro A, Tanimoto K, Matsuoka Y, et al. Degos disease with constrictive pericarditis: a case report. Jpn Circ J. 2000;64:464-7.
Figures

Figure 1

Computed tomography. Computed tomography demonstrates bilateral pleural effusion and pericardial effusion with marked calcification of the pericardium.

[5] Nomland R, Layton JM. Malignant papulosus with atrophy (Degos); fatal cutaneointestinal syndrome. Arch Dermatol. 1960;81:181-8.
**Figure 2**

Cardiac catheterization. Cardiac catheterization demonstrates equal right and left ventricular end-diastolic pressures and square root signs. SVC, superior vena cava; IVC, inferior vena cava; RA, right atrium; RV, right ventricle; rtPA, right Pumonary artery; rtPCW, right pulmonary capillary wedge pressure; LV, Left ventricle; Ao, Aorta

|            | SVC | IVC | RA | RV | rtPA | rtPCW | LV   | Ao  |
|------------|-----|-----|----|----|------|-------|------|-----|
| Systolic/diastolic (mean) pressure mmHg | 16/16 (14) | 14/15 (15) | 16/16 (15) | 39/10 (9) | 36/21 (26) | 19/19 (18) | 116/10 (20) | 107/61 (80) |

**Figure 3**

Intraoperative gross findings. (A) The inside of the pericardial sac displays adhesions with some calcification. (B) A high degree of calcification in the myocardium.

**Figure 4**

Histopathological findings. (A) There was a high degree of calcification and fibrosis in the pericardium (green arrow). (B) Lymphocytic infiltration around the pericardial vessels (blue arrow).