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Case Report

Dego`S Disease with Pleural Effusion. Case Report and Review of Literature

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Abstract

Degos' disease, also known as "malignant atrophic papulosis" is a rare vasculopathy characterized by typical cutaneous lesions with an unknown etiology wich was first described by Dego in 1942 [1], but another case, reported in 1941 by Köhlmeier, who interpreted it as thromboangiitis obliterans of the mesenteric vessels [2]

This is an arteriopathy which involves vessels of small-caliber. Dego disease mostly affects the skin. It occurs as a limited benign, cutaneous form and in a systemic variant potentially lethal multiorgan. Male to female ratio of this disease is (3:1), and a few cases of familial involvement have been reported [3-7]. In 60% of reported cases there has been an involvement of the gastrointestinal tract and other organs [8]. Up to now, fewer than 50 living patients are known worldwide, and fewer than 200 cases reported in medical literature, because it is underdiagnosed due to the rarity of this nosology [9,10]. Patients shows symptoms between of 20- 50 years; however, symtoms have been reported even as 8 months [1,6].

Keywords: Malignant Atrophic Papulosis (MAP); Pleural Effusion; Dego

Case Report

A 58 -year-old man presented to the hospital with productive cough, dyspnea, chest wall pain,and in chest x-ray examination was observed left pleural effusion.

Five months ago, the patient diagnosed with Dego disease, presented by more than 50 remittent eruptions which were surrounded by borders erythematico - teleangiectatic, varying from 5-8 mm in diameter, which were located mostly on trunk antero-posterior position, that started one year ago without the complete disappearance of older ones (Figure 1 and 2).

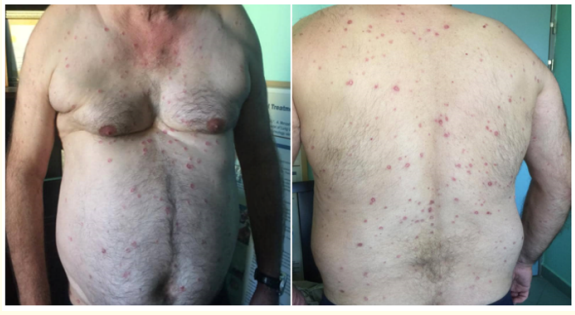

Figure 1: Skin photograph showing white to pink papules, 5–10mm in diameter, with central, porcelain-white atrophic center surrounded by a peripheral telangiectatic rim.

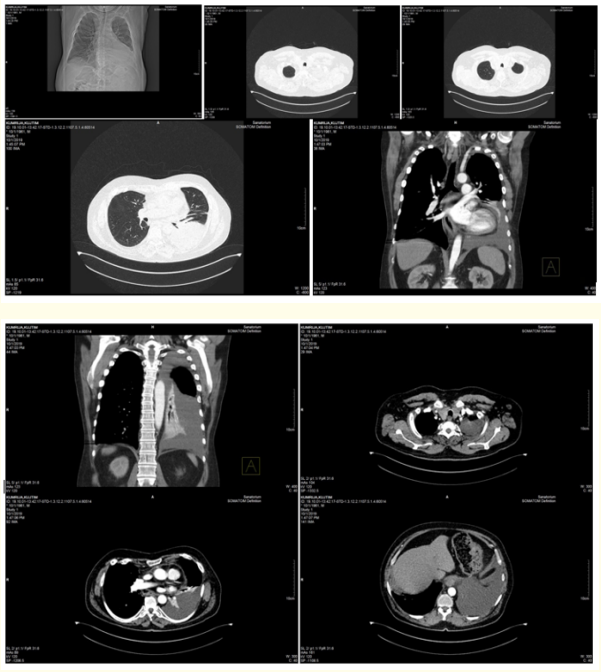

Figure 2: Left pleural sapce pleural effussion.

The eruptions were painless and nonpruritic.

The patient was also taking medication for hypertension.

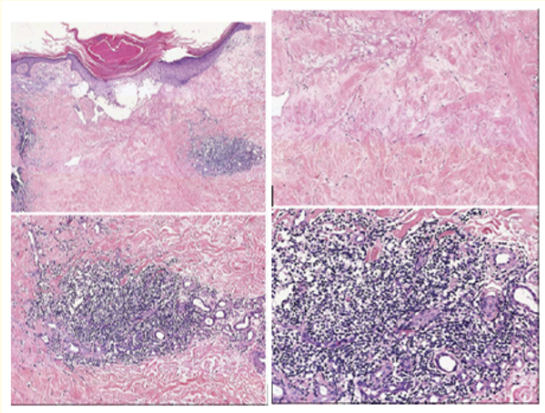

Figure 3: Histopathology of the skin biopsy by hematoxylin and eosin staining :Epidermal atrophy, hyperkeratosis, and wedge-shaped dermoepidermal necrosis were visible the panels demonstrated occlusion of blood vessels in the corium and perivascular lymphocytic infiltration.

During the hospitalisation were performed a series of examination to find the cause of pleural fluid. A therapeutic thoracentesis was performed and 1L of green exudative fluid was removed with pH 7.26, WBC 260 cells/mm3 (PMN 16%), LDH 746 IU/L, triglycerides 30 mg/dL, and total protein 5.2 g/dL, cholesterol 60mg/dl,neutrofils 3%, lymphocytes 27%, macrophages 70%. G-expert was negative for TB infection.

Other analysis results were: eritrocytes sedimentation 7,fibrinogen 442,PT 13.5, PT/INR1,APTT 28.5,CRP 2.9.

Hepatic panel was normal, mineral panel normal, LDH 230,BNP 54,total protein 7.9,RPR nonactive, uranalysis wit microscopy was normal.

In the condition of the persistence of pleural fluid even with the wide specter of antiobioticotherapy was decided, in the multidisciplinary team for diagnostic and therapeutic VATS (Video Assisted Thoracoscopy).

With lidocaine 2%- 40 ml diluated, in 4-5-th space, lineaaxillaris anterior and media,we opened pleural cavity, aspirated the pleural fluid and taken some pieces from parietal pleura for histopathological examination

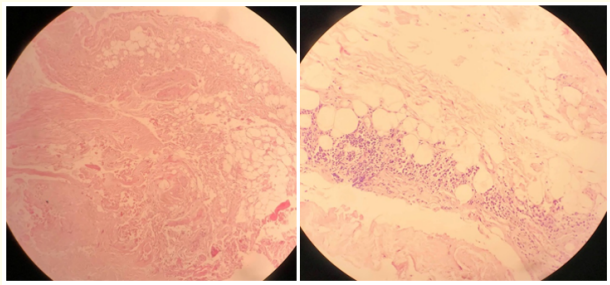

Figure 4: Histopathology of parietal pleuara showed fibrosis and perivascular limfocitic infiltrate (H-Ex10, H-E x40).

The patients was in good condition after VATS with medication. Pleural biopsy resulted with fibrosis and chronic inflammation perivascular limphocyti cmononuclears, no granulomatous process, wich reveals a systemic Dego disease.

Discussion

Degos' disease, is a rare condition with skin lesions on the trunk and proximal portion of the extremities, but soles, face and palms tend to be spared, like our case, based on largely papules, surrounded erythema and telangiectasias.

Only in 37% of patients, has been reported purely cutaneous form [11].

It occurs as a limited benign, cutaneous form and in a systemic variant potentially lethal multiorgan [12,13].

Endothelial proliferation and swelling of small and medium size blodd vessels are the main features of this disease.

The etiology of the disease remains still unclear. Some believe that Degos' disease is a thrombotic rather than an autoimmune disease, because no circulating immunocomplexes, ant endothelial cell antibodies, or anticardiolipin antibodies are isolated, even there a a few cases, where antiphospholipid antibodies are identified.

The other interal organs affected, according to the frequency are: gastrointestinal tract, central nervous system, thoracic organs and kidney.

Approximately in 60% of reported cases, the involvement of gastrointestinal tract is presented (from the oral cavity to the anus) [3], with the predominance of small bowel [9] and in the condition of the intestinal perforation, occurs death of the patient [6].

Neurologic manifestations occur in around 19% of patients, including cerebral hemorrhage, subdural hematoma, thrombosis of cerebral arteries, venous sinus thrombosis, encephalitis, meningitis, polyradiculoneuropathy, cranial neuropathy, and myopathy [14,15].

There are very few cases reported in the medical literature with pleural involvement of Dego disease. Pierce and Smith described a case where the patient died three months after decortication of the left lung, due to respiratory disjunction and heart failure [16].

Our patient had no gastrointestinal, or neurological findings and based on literature review, we recommend medical treatment for MAP, whether cutaneous or systemic form, remains to be defined.

Antiplatelet drugs plays an important role in the treatment of all variants of MAP [9]. We used Aspirin and Dipyridamole for our patient, improving his general condition.

Conclusion

The features of our case highlight the importance of considering systemic Degus disease in case of pleural effusion especially when there is skin involvement.

Skin biopsy is essential for the diagnosis of this entity. Platelet antiaggregant as well as anticoagulants seemed effective to the control of the disease.

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None.

Conflicts of Interest

There are no conflicts of interest.

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