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Case Report

Bowel perforation in Dego's disease: a lethal surgical scenario

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ABSTRACT

Malignant atrophic papulosis or Dego's disease is a type of cutaneous disease with involvement of the gastrointestinal and central nervous system. Involvement of the gastrointestinal system by way of perforation invariably leads to a fatal outcome. Awareness of this condition will help in providing a high index of suspicion while managing unexplained multiple intestinal perforations or while dealing with rapidly developing complications in perforative peritonitis.

Keywords: Kohlmeier, Dego's disease, Manifestation, Treatment

INTRODUCTION

Dego's disease also described as malignant atrophic papulosis (MAP) is an extremely uncommon condition of the skin associated with multisystem morbid complications. The condition was first described by KohlMeier in 1941 and later documented as a separate entity by Robert Degos.[1] The patho- physiology of the disease continues to be obscure with less than 200 cases reported in world literature. Since multisystem involvement of the disease more so of the gastrointestinal tract is associated with high mortality, awareness of this condition is extremely important for a general surgeon.

A case of Dego's disease manifesting as severe perforative peritonitis is reported along with a brief review of literature.

CASE REPORT

A 27 year old male presented with history of severe pain in abdomen of 4 hours duration accompanied with vomiting and distention of abdomen. He sought treatment from his family physician. But as the severity of the symptoms increased, he was referred to our surgical unit. On examination he was febrile with a pulse of 110 /min and a B.P of 110/70 mm of Hg. Patient was dehydrated but didn't have icterus. Examination of the trunk revealed multiple white cutaneous lesions which were papillary in nature (Figure 1). Per abdominal examination revealed board like rigidity. Genitalia were unremarkable. Patient was administered aggressive resuscitation. Laboratory investigations revealed neutrophilic leukocytosis. Rest of the laboratory reports including coagulation profile were within the normal range. Chest x ray revealed free gas under the diaphragm. Ultrasound of the abdomen revealed distended bowel loops and significant amount of free fluid. Patient underwent exploratory laparotomy. At laparotomy there was around 2 liters of purulent fluid and a perforation in the small bowel approximately 1.5 feet distal to the duodenojejunal junction. There was another lesion approximately 1 foot proximal to the IC junction, which showed excessive thinning of the bowel wall .The edges of the perforation were freshened and sutured. For the serosal lesions reinforcing interrupted seromuscular sutures were taken. Drains were placed in the peritoneal

cavity. Drain output was serous and negligible by the fourth postoperative day. Biopsy of the cutaneous lesions revealed changes typical of Dego's disease (Figure 2). On the fourth postoperative day the patient suddenly became breathless and hypotensive. ABG showed severe metabolic acidosis. Repeat chest x ray did not reveal any abnormality. Patient was rein-tubated and kept on ventilatory support. Inotropic support was also commenced to maintain the blood pressure .The abdomen did not reveal any abnormality as the abdominal girth was well maintained and the drain output was all most negligible. The patient was kept on ventilatory support for almost 3 days after which he expired.

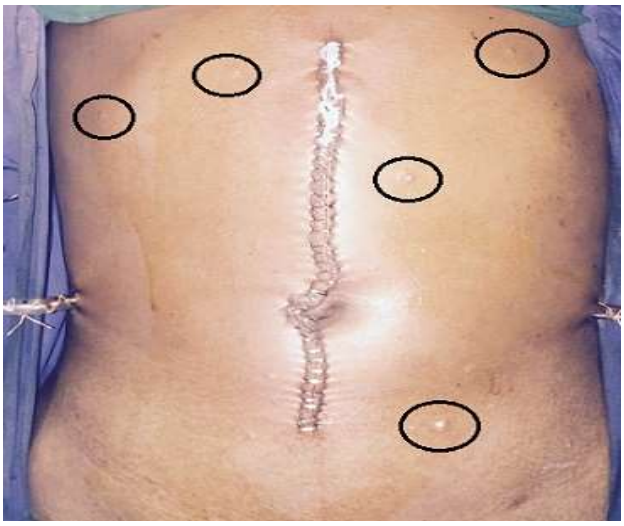

Figure 1: Typical lesions seen on the trunk marked by black circles.

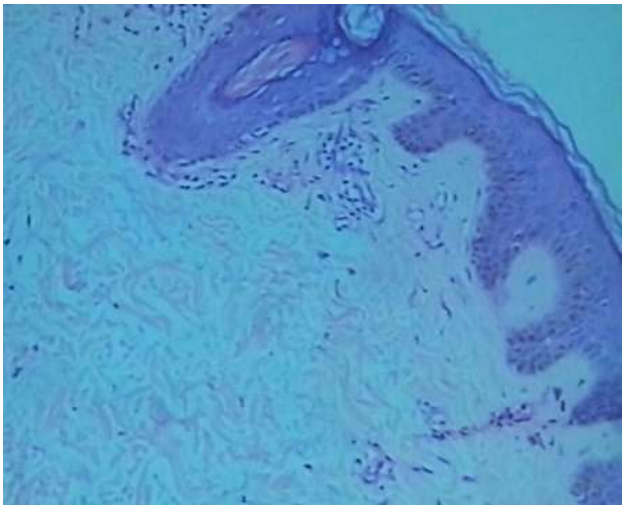

Figure 2: Histology of the cutaneous lesion revealed hyperkeratosis and keratin horns with finger like elongations of rete ridges. There was increased melanin pigmentation in the basal layer of the epidermis with perivascular chronic inflammatory infiltrate in the dermis. (H& E staining, Magnification 40X).

DISCUSSION

Malignant Atrophic Papulosis is a very rare disease primarily affecting the skin, but the sequel of this is commonly seen in other organ systems. There are less than 200 cases reported in literature making this disease a rare entity. Therefore there is a need to report this case.[1,2] The cutaneous manifestations are in the form of multiple papules with white center surrounded by erythematous telangiectatic borders. The size may vary from 5 to 10 mms in diameters (Figure 1). They are non-pruritic and painless with predilection for the trunk and extremities.

In addition to the skin, the gastrointestinal system is commonly affected manifesting as multiple perforations of the intestine. The other organ systems affected are the central nervous system and the kidneys in a few cases.[4,5] However the prognosis of patients of Dego's disease presenting with perforative peritonitis is extremely poor as seen in the case presented. This renders awareness of the condition and its accompaniments mandatory. The pathophysiology of this disease is quite complex with no well-defined pathognomonic features. Three hypotheses have been postulated to explain the pathophysiology.

a. Inflammation of the vessels could act as a trigger factor for the development of MAP.[4,5] The vasculitis factor bears close similarity to SLE. Deposits of C5B9 have been found in the skin, GI tract and brain vessels in patients suffering from MAP. Majority of these cases have evidence of high expression of alpha interferon, endothelial tubercular reticular inclusions and interferon gene expression in the peripheral blood mono nuclear cells.

b. The next hypothesis postulates Dego's disease as a coagulopathy wherein the thrombus in the vessels of the dermis is the initiating event in MAP. The reduction of blood flow with the resulting damage of the endothelial cells leads to deposits of mucin and aggregation of mono nuclear cells. The other associated findings may be fibrinolytic dysfunction and alteration in platelet aggregation.[5]

c. MAP can be a fall out of primary or secondary dysfunction of endothelial cells with abnormal swelling and proliferation of the vascular endothelium which could trigger thrombosis of cutaneous, intestinal and brain vessels. Therefore a viral infection can be postulated to act as a cause for endothelial changes.[6]

Since the pathophysiology of the disease is obscure, variable management strategies have failed to control the disease. Medical treatment in the form of anti-platelet medications such as aspirin, dipyridamole etc. can be administered assuming the disease to be a type of vasculitis.[3] Immunosuppressive therapy have been tried out with very poor results .Therefore anti platelet agents still continue to be the first line agents for this condition.

Treprostinil which is a prostaglandin analogue has been tried in a few patients with sub optimal results.

Majority of the patients suffering with MAP manifest with features of perforative peritonitis.[7,8] Since the pathology involves medium to small size vessels of the gut, the disease could be wide spread at the time of the presentation. Multiple intestinal perforations is usually the commonest presentation. If one is unaware of this rare condition, the surgeon could be bewildered while busy searching for the etiology of multiple perforations. It is only awareness of the cutaneous manifestations of the disease or histological confirmation of the biopsy specimen obtained from the skin lesion that leads to an established diagnosis.

The mortality and morbidity associated with intestinal perforations in patients with this condition is extremely high.[9,10] Awareness of this phenomena is pivotal for the attending surgeon. This is because if the cutaneous lesions are undiagnosed the cause for high mortality may go unrecognized. Therefore if confronted with multiple intestinal perforations which do not fit into the diagnostic criteria of commonly encountered GI diseases, one needs to look for cutaneous lesions which could go in a definitive way to arrive at a tentative diagnosis. The poor prognosis in patients with established diagnosis of the disease has to be explained prior to surgical intervention.

CONCLUSION

Dego's disease or MAP is an extremely rare cutaneous disease with obscure pathology. Gastrointestinal complications in form of perforation are very common in this disease.

They have very high morbidity and mortality. For patients who survive surgical intervention for perforative peritonitis the only modality of treatment available is anti-platelet therapy.

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