

Malignant Atrophic Papulosis

A Agarwal, K Shivaswamy, D Thappa, S Jayanthi

Citation

A Agarwal, K Shivaswamy, D Thappa, S Jayanthi. *Malignant Atrophic Papulosis*. The Internet Journal of Dermatology. 2006 Volume 5 Number 2.

Abstract

Sir,

A 21 –year- old male presented to our dermatology OPD with multiple asymptomatic raised skin lesions on the trunk, neck, arms and forearms of six months duration. The patient had been referred to us from Surgery department for evaluation. He had undergone an abdominal surgery (a month ago) for multiple ileal perforations for which an ileal resection was done. He had no other systemic complaints. There was no family history of similar illness.

Patient appeared cachectic, vitals were normal and cutaneous examination revealed multiple, scattered, discrete erythematous papules with telangiectetic borders of varying size ranging from 0.5 cm to 1 cm over the anterior chest (Fig. 1), abdomen, neck, arms and forearms.

Figure 1

Figure 1: Multiple, scattered, discrete erythematous papules with telangiectetic borders over the chest

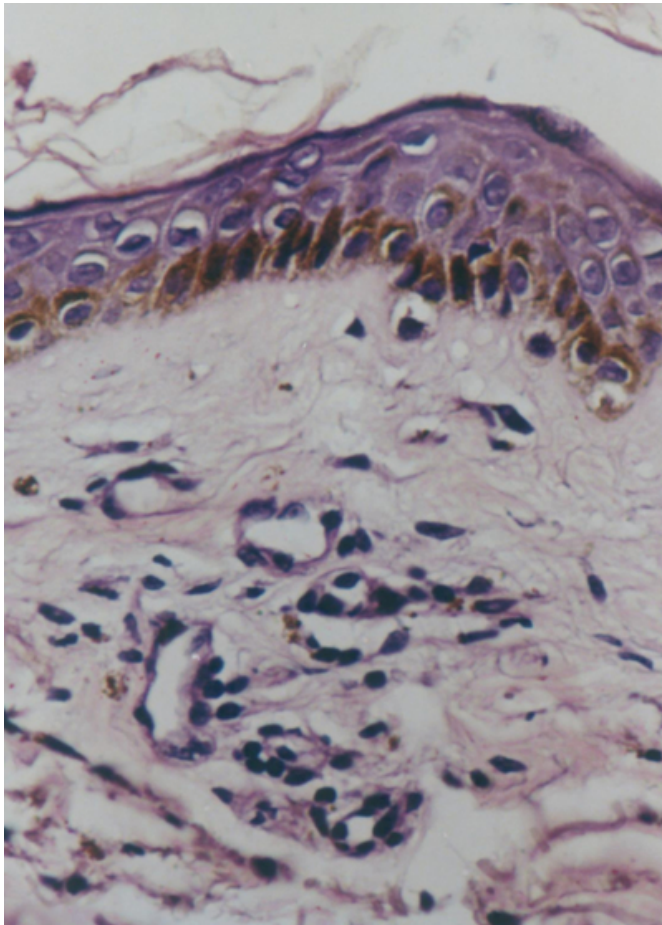


Some lesions over the neck and chest showed central porcelain white atrophy. The bulbar conjunctiva showed an avascular patch surrounded by collateral vessels. Oral and genital mucosa was normal. A clinical diagnosis of

malignant atrophic papulosis was made. Routine investigations including CBC, ESR, liver and renal function tests were within normal limits. Tests for collagen profile including antinuclear antibody were negative. VDRL and ELISA for HIV were negative. Histopathological examination from one of the papules revealed an atrophic epidermis with a zone of underlying acellular collagen and dermal capillaries with endothelial cell swellings with abundance of mucin among collagen bundles (Fig. 2). These findings were consistent with our clinical diagnosis of malignant atrophic papulosis.

Figure 2

Figure 2: Higher magnification photomicrograph demonstrating atrophic epidermis and capillaries with endothelial cell swelling (Haematoxylin & Eosin X 400)



Malignant atrophic papulosis, also known as Degos disease was first described by Kohlmeier in 1941 and subsequently by Degos, Delort and Triscot in 1942.¹ It is a rare, often fatal, multisystem disorder in which pathognomonic skin lesions are frequently associated with infarctive lesions of other viscera, particularly the gastrointestinal tract.² Some patients have only cutaneous lesions and a relatively benign course.³

The aetiopathogenesis of this syndrome is unknown. Degos disease is vaso-occlusive in origin. It has been regarded as lymphocyte mediated endovasculitis or primary endothelial defect with secondary thrombosis leading to infarctive lesions.⁴ Few reports emphasize the possibility of genetic transmission of this disease.¹ Slow virus infection and environmental factors have also been proposed.¹

A male predominance has been reported. The age of the onset is usually between 20 and 40 years. In this disorder, the skin and viscera are involved with focal infarctions

caused by vasoocclusion. The organ most involved is the skin. Skin lesions are usually the first sign of the disease. They present initially as pink or red dome shaped papules of 2-15mm in size. Some of the lesions persist as papules and a few disappear rapidly leaving a tiny white scar. The majority, however, rapidly develop a central area of necrosis which replaces the papule with a central scale of grayish white colour which has been exactly likened to porcelain.^{5, 6} This atrophic centre is ringed with telangiectasia. The lesions occur predominantly on the trunk and upper limbs. The palms, soles and face are usually spared and the scalp is not affected. Similar lesions in the gut give rise initially to abdominal cramps, vomiting and enteritis. These symptoms usually manifest themselves from 3 weeks to 3 years after the appearance of skin papules, but occasionally precede these. Sooner or later they are replaced by an acute, terminal intestinal crisis due to haematemesis or perforation leading to peritonitis as seen in our case. Mucous membrane involvement is very rare and when it is seen it is usually ocular, in the form of avascular patches over the bulbar conjunctiva surrounded by collateral vessels as evident in our case. Sclera, episclera, retina, choroid and optic nerve may be involved very rarely. When the nervous system is involved the cerebral infarctions manifest as headache, transient neurological defects, multi-infarct dementia and epilepsy. The pericardium, myocardium, pleura, lungs, liver, pancreas and genitourinary tract also may be involved.^{1, 5, 6}

The histological appearance of malignant atrophic papulosis is typically characterized by a wedge-shaped area of necrosis (altered dermis) from the epidermis through the dermis. Beneath the zone of ischemia, there is endothelial swelling and proliferation; thrombosis may variably be seen. There may be a sparse perivascular mononuclear cell infiltrate, but there is no vasculitis. More common, however are edema, extensive mucin deposition and slight sclerosis. Initially, the mucin deposits are localized to the ischemic zone, but in older lesions the material is confined to the margins of this zone. They stain with colloidal iron or alcian blue.²

The characteristic features of malignant atrophic papulosis can rarely be confused. A similar cutaneous – intestinal syndrome (having combination of macular, blistering and crusting lesions of the skin with oropharyngeal ulceration and death from perforation of many intestinal ulcers) does not show the characteristic histology. Identical lesions have been described in cases of rheumatoid arthritis, lupus erythematosus and scleroderma and one case of Crohn's disease.⁵

There is no effective treatment, although phenylbutazone was thought to have caused remission. Steroids do not help, and even identification and removal of the perforating intestinal plaque (which is exceedingly difficult) only leads to temporary respite. Aspirin, dipyridamole, prostaglandin E1 and fibrinolytic therapy have also been advocated.^{5, 6}

Classically, the prognosis is poor and the treatment does not modify the prognosis of the disease. Death ensues most often after intestinal perforation and peritonitis, sometimes after nervous system involvement, or as part of the picture of ill-defined respiratory insufficiency. Deaths may exceptionally be attributed to renal infarct and pulmonary abscess. However, the survival varies from months to years.⁶

CORRESPONDENCE TO

Dr. Devinder Mohan Thappa, MD, DHA, MNAMS,
Professor and Head, Department of Dermatology and STD,

JIPMER, Pondicherry - 605 006, India Email-
dmthappa@gmail.com; dmthappa@vsnl.net

References

1. Katz SK, Mudd LJ, Roenigk HH. Malignant atrophic papulosis (Degos'disease) involving three generations of a family. *J Am Acad Dermatol* 1997;37:480-484.
2. Weedon D. *Skin Pathology*, 2nd edn. London: Churchill-Livingstone, 2002:221-278.
3. Farrel AM, Moss J, Costello C, Fearfield LA, Woodrow D, Bunker CB. Benign cutaneous Degos' disease. *Br J Dermatol* 1998; 139:708-712.
4. Tribble K, archer ME, Jorizzo JL, Sanchez R, Solomon AR Jr, Gardner FH, Cavallo T. Malignant atrophic papulosis: absence of circulating immune complexes or vasculitis. *J Am Acad Dermatol* 1986; 15:365-369.
5. Ryan TJ. Cutaneous vasculitis. In: Champion RH, Burton JL, Burns DA, Breathnach SM, editors *Rook/Wilkinson/Ebling Text Book of Dermatology*., 6th edn. Oxford-Blackwell science, London. 1998; 2155-2225.
6. Degos R. Malignant atrophic papulosis. *Br J Dermatol* 1979; 134:231-236.

Author Information

Anuja Agarwal, M.D.

Ex Senior Resident, Department of Dermatology and STD, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER)

K. N. Shivaswamy, MD, DNB

Senior Resident, Department of Dermatology and STD, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER)

Devinder M. Thappa, MD, DHA, MNAMS

Professor and Head, Department of Dermatology and STD, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER)

S. Jayanthi, MD

Professor and Head, Department of Pathology, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER)