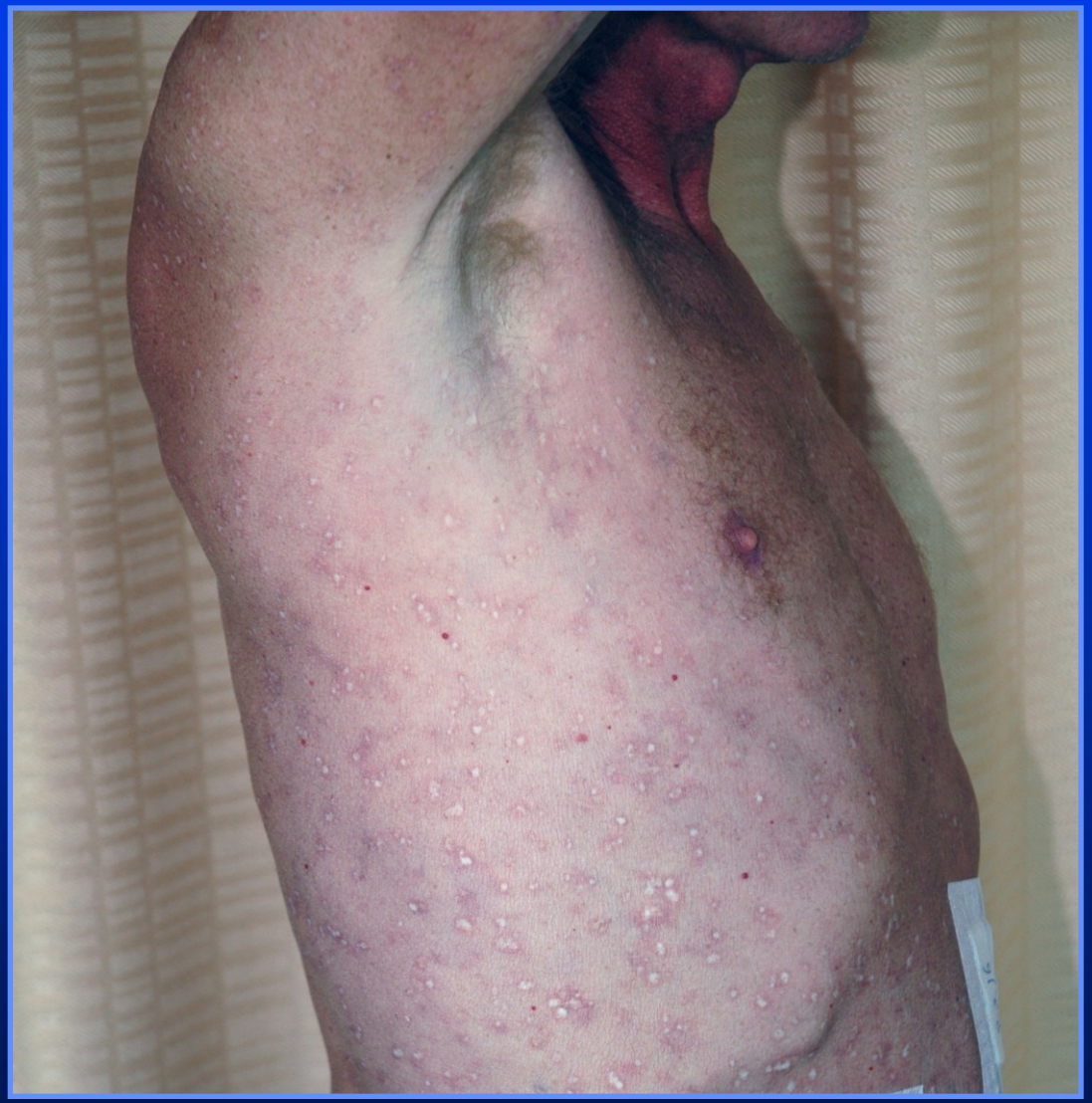
A 60 year-old male is admitted with sudden left visual loss.

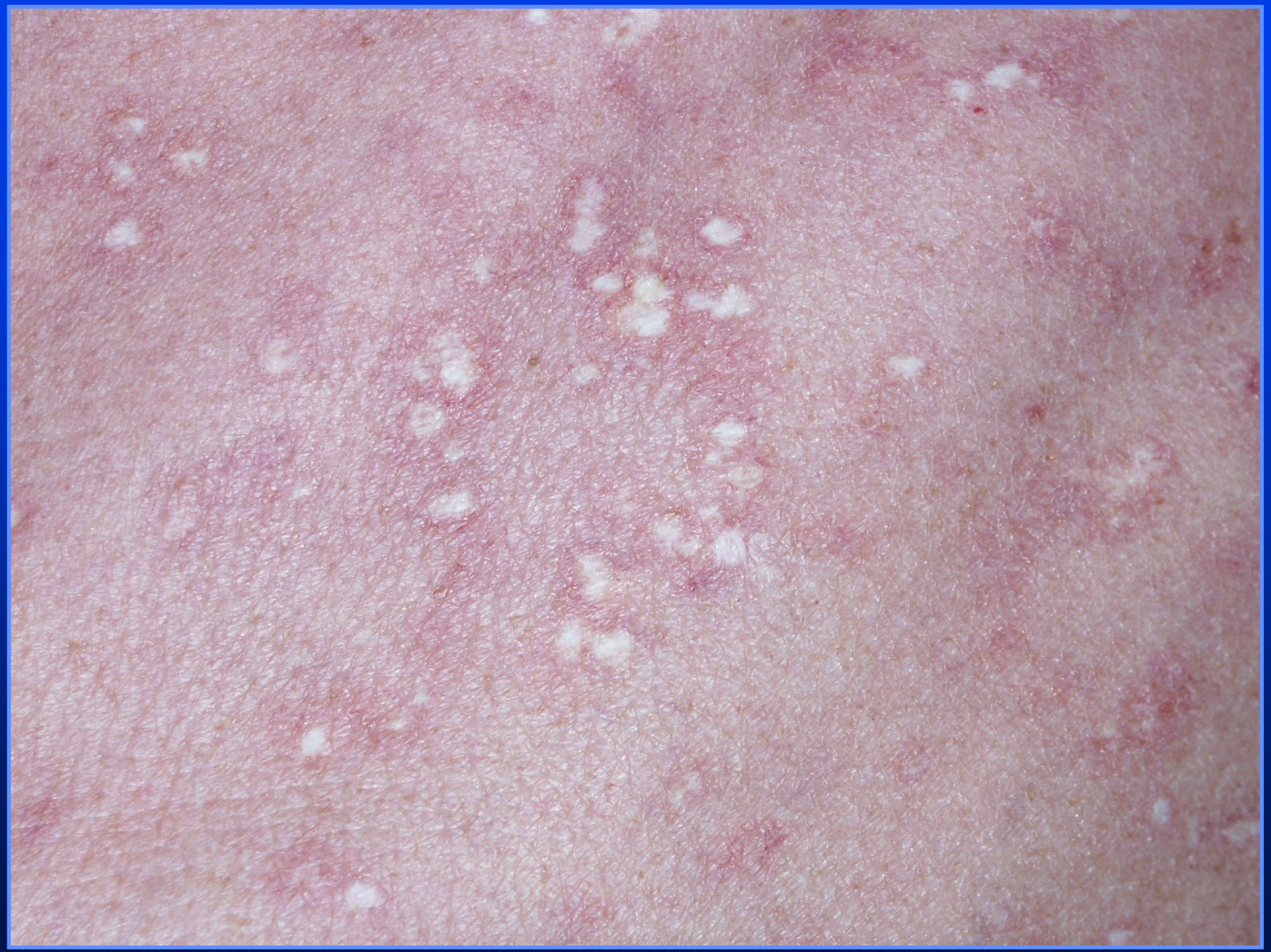
He had complained for 18 months of post-prandial abdominal cramps and diarrhoea but investigations had been inconclusive. He lost 10 kilos of weight. A diagnosis of functional colopathy was done. Since then dysphagia and important weakness have appeared.

He also evoked cutaneous lesions on the trunk and proximal limbs, which had progressively developed over the last 3 years. A skin biopsy was performed and showed aspecific lymphocytic vasculitis.

Physical examination

shows left optic papillary oedema, global amyotrophy and symmetrical lower limb paresis, hypoesthesia of the left crural nerve territory and a diffuse cutaneous cicatricial papulosis (central scars surrounded by an erythematous border) on the trunk and proximal limbs.





- The *laboratory tests* show neither autoimmune nor inflammatory abnormality.
- *Intestinal angio-MRI* (Magnetic Resonance Imaging) is normal.
- *Triolein breath test* shows severe malabsorption.
- *Oesogastroduodenoscopy and small gut videocapsule* show a diffuse congestive mucosa.
- *Optic fundus* reveals left ischaemic optic neuritis.
- *Cerebral MRI* shows bi-hemispheric multiple small infarcts.

Degos disease

- Degos disease or malignant atrophic papulosis is a rare vasculopathy characterized by narrowing and occlusion of the lumen by intimal proliferation and thrombosis, which leads to ischemia and infarction in the involved organ systems.
- 150 cases have been described in the literature. It affects 3 males for 1 female and has been associated with systemic lupus, antiphospholipid antibody syndrome, HIV, or described in children.

Degos disease

- A benign cutaneous form exists (37%) and has a very good prognosis.
- The systemic form (63%) is always fatal, death is due by abdominal, neurological, pleural or pericardial infarction. The mean life expectancy after diagnosis is 1 to 2 years.

Degos disease, systemic form

- Digestive tract lesions: 50%
  - With abdominal cramps, malabsorption, weight loss and diarrhoea
- Ocular lesions : 35/105 cases described
  - With optic neuritis, cataracts, optic nerve atrophy, oculomotor nerve palsy
- Neurological lesions
  - With cerebral ischaemic or haemorrhagic stroke, medullar strokes, radiculitis, headache, epilepsy
- Pleural or pericardial involvement with effusion
- All other organs can be involved…

Degos disease

- Physiopathology is still unknown. Thrombotic, immunological and viral hypotheses have been proposed.
- No effective treatment exists, while some drugs seem to delay the occurrence of the lesions. The most used are aspirin and pentoxyfillin, and cortisone.

  Our patient received oral cortisone, clopidogrel, and intravenous immunoglobulins, but he developed cerebral transient stroke and pleural effusion, and the digestive malabsorption didn't get better. Then last month he received infliximab, an anti-TNF drug.

Degos disease

Conclusion

Degos disease is a clinical entity which associates pathognomonic cutaneous papulosis and ischemic organic lesions. Its rarity and rapidly fatal course make the disease a difficult diagnostic and therapeutic challenge.

References

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