The pathogenesis of the sialadenitis is not certain. Bock and Wright (1964) have shown that genetic factors are capable of causing much of the variability which is seen in response to iodine administration. Follis (1961) has studied iodide-induced submandibular sialadenitis in the hamster. Injection into the animals of iodide in daily doses equivalent to the 6 g. taken by the patient in the present study resulted in damage to the distal duct system. He concluded that the inflammation was a direct toxic effect of iodide on the cells.

In man, as in the hamster, iodide is concentrated in saliva to many times the plasma level (Harden *et al.*, 1966), and there is evidence to suggest that the site of concentration is the salivary ducts (Mason *et al.*, 1966). Acute enlargement of the thyroid has also been recorded as a toxic effect of iodide, with rapid onset and rapid subsidence when iodide was stopped (Edmunds, 1955). A critical amount of iodide must be administered to produce this change in the thyroid, and a high concentration of iodine must be reached in the gland if the change is to occur (Follis, 1961). The stomach and the breasts also concentrate iodine. The stomach concentration of iodide may explain some of the gastrointestinal symptoms seen in iodine overdosage. The breast concentration may explain the failure of lactation in rats fed at high levels of inorganic iodine reported by Ammerman *et al.* (1964) and Arrington *et al.* (1965).

Degos's Syndrome

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Degos *et al.* (1942) described a disorder characterized by a papular skin eruption progressing to ulceration and followed by a fatal intestinal episode. The name malignant atrophic papulosis was coined to emphasize the fatal outcome. Since then 13 cases have been reported. A further case is here recorded and the literature reviewed.

CASE REPORT

A 26-year-old man was admitted as an emergency on 29 March 1966 with a 24-hour history of malaise and fever. Coincident with the onset of the illness he developed a number of skin lesions affecting principally his limbs, face, and to a less extent his trunk. In the past he had been fit and had not taken any drugs. On examination he looked ill and was febrile. The skin lesions were tender papules 0.25-1 cm. across, and the larger ones had already developed a black centre (Fig. 1). No abnormality could be found in the chest or abdomen.

Investigations.—Hb 15.2 g./100 ml. W.B.C. 22,300 (78% P., 14% L., 8% M.). E.S.R. 11 mm. per hour. Urine N.A.D. Brucella and salmonella agglutinins negative, serum proteins 7.1 g./100 ml. (albumin 2.75 g., globulin α_1 0.94 g., α_2 1.04 g., β 0.94 g., γ 1.43 g.) Electrolytes and urea N.A.D., W.R. negative. Antinuclear factor not detected. Latex fixation non-reactive. Calcium, phosphate, and alkaline phosphatase normal. Chest x-ray examination N.A.D.

The provisional diagnosis on admission was polyarteritis nodosa. A skin lesion was biopsied and prednisone 10 mg. t.d.s. was given. There was a subjective improvement after 24 hours, but the skin lesions progressed and became necrotic in the centre (Fig. 2). He also developed ulcers on the buccal mucosa and lower lip. On the eleventh day of illness he complained of abdominal pain and nausea, and a surgical opinion was sought. On examination at that time he was apprehensive, dehydrated, and in pain. The abdomen was distended, tender, and silent; generalized peritonitis was diagnosed, possibly due to a visceral complication of polyarteritis. At laparotomy a perforation of the ileum was found about 4 ft. (1.2 m.) from the ileocaecal valve. Over an area of 10 in. (25 cm.) in the

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region of the perforation the bowel was studded with small oval white subserous plaques; these corresponded to areas of mucosal ulceration. The affected piece of bowel was resected. There was a similar yellow plaque on an adjacent piece of sigmoid colon, but no thinning of the wall, and it was felt that this was serosal reaction secondary to the adjacent small-bowel perforation. The abdomen was closed with a drain to the region of the sigmoid colon.

FIG. 1

The postoperative course was satisfactory until the fourth day, when he suddenly developed abdominal pain and began to discharge faecal material from the drain. A further laparotomy was performed, the wound showed no sign of healing and was full of pus. The small-bowel anastomosis had fallen apart, while the descending and sigmoid colon was gangrenous. The principal mesenteric vessels were pulsating normally. The proximal end of the ileum was brought out as an ileostomy as it was felt that the fashioning of any further anastomosis would be fraught with danger. The necrotic colon was excised and the ends of the bowel were exteriorized. Postoperatively the patient remained extremely ill;

his skin lesions progressed to frank ulcers but no fresh lesions appeared. At this time he was seen by the consultant dermatologist, Dr. J. D. Everall, who made a diagnosis of Degos's syndrome. The patient was by now off corticosteroids, there were difficulties in controlling his ileostomy, and his wound dehiscd, but despite this he seemed to hold his own for two months. By July many of the skin lesions had healed, leaving a characteristic depressed atrophic scar. Despite various intravenous supplements his general state of nutrition remained poor, he developed a lung abscess and empyema, and died on 26 September, six months after the start of his illness.

The histological appearances of the original skin biopsy and the small and large gut removed at the two operations are described below.

(1) *Skin Biopsy*.—Sections show ulceration with oedema and haemorrhage into the dermis. Some polymorphs are seen, but there

FIG. 2

is a paucity of inflammatory cells. The vessels show swelling of the endothelium with in some cases apparent proliferation. The main vascular changes involve the intima almost exclusively where in some cases fibrinoid material may be demonstrated.

(2) *Gut*.—Both specimens show areas of mucosal necrosis and ulceration; there is a marked exudate but relatively little polymorph reaction. In both specimens there was marked oedema and vascular dilatation in all layers. The blood vessels, both arteries and veins, show changes varying from swelling of the endothelium almost amounting to proliferation to, in some cases, frank fibrinoid degeneration. In some blood vessels thrombosis was seen. The main impact of the disease process seemed to be on the intima, the remainder of the vessel wall apparently escaping injury.

COMMENT

Despite the title of this syndrome the first person to describe the condition was Köhlmeier (1940). His patient presented with small necrotic skin lesions and died from similar lesions affecting the bowel. Since then some 13 cases have been described and from a review of the literature a definite clinical picture emerges. The disease affects predominantly males usually in their third decade. The skin lesions are characteristic and affect principally the trunk, proximal extremities, and neck; the face is occasionally involved as in the above case. The palms and soles are spared, as are the mucous membranes, though again in this patient the buccal mucosa and lips were involved. The initial lesion is a barely raised erythematous papule 2–5 mm. across; as the papules enlarge they often lose their circular outline and take on irregular forms, sometimes

linear or rectangular. Gradually over days or weeks they become umbilicated and develop a central scale which falls off, leaving a depressed central atrophic scar. The cutaneous phase may take three weeks, as in the above case, seven months (Degos *et al.*, 1942), one year (Köhlmeier, 1941), or three years (Lausecker, 1949), during which successive crops of papules may make their appearance. At any time there may be all stages of the lesion.

In the above case the abdominal symptoms occurred after only 10 days, but more often some weeks or months elapse before the abdominal phase of the syndrome is encountered. The symptoms and signs are those of peritonitis, and at laparotomy the small bowel is studded with white subserous plaques corresponding to shallow ulcers on the mucosal aspect. Perforation of one of these as in this case is responsible for the peritonitis. Lesions may also occur in the stomach (Lausecker, 1949) or, also as in the above case, the colon (Basset *et al.*, 1956; Naylor *et al.*, 1960; Hall-Smith, 1964). Peritonitis necessitates exploratory laparotomy but has proved beneficial in only one case. The impressive thing in our patient was the complete absence of healing in the wound or abdominal cavity at the second laparotomy. This must have been due to the disease process, as corticosteroids alone in the dosage used would not have had this disastrous effect. In previous cases corticosteroids were tried, but when intestinal symptoms developed the course was steadily downhill and all therapy useless. It is of some interest that two patients developed the classical skin lesions but did not then apparently develop manifest intestinal lesions (Ingram and Sutherland, 1957; Howell, 1957); both these patients survived.

The cause of this condition remains unknown. It has been suggested that it is a clinical variant of polyarteritis nodosa; but while age and sex would fit, the pathological process appears to be different. In Degos's syndrome, regardless of the site, the pathological changes are similar and seem to be mainly vascular; the other changes are probably secondary. Small blood vessels, both arteries and veins, are involved and the inflammatory process seems to be maximal towards the intimal surface. The elastica and media are scarcely involved. Endothelial proliferation may be seen, and in some areas fibrinoid degeneration. Where the blood vessels are maximally involved there may be thrombosis. In polyarteritis, however, the basic lesion is a panarteritis involving somewhat large arteries. Necrosis of the media and intima, including the elastic lamina, is seen. From the histological appearances, Degos's syndrome is undoubtedly a vasculitis which may in the future be attributed to an immune or autoimmune process.

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