Meeting March 19 1964

Cases

Malignant Atrophic Papulosis (Degos's Syndrome)

S P Hall-Smith MD FRCPEd

T L, male, aged 16

History: In September 1963 developed generalized eruption which achieved its present widespread extent within a week or two. At that time he felt well in himself and was able to continue with his farm work. At Christmas consulted his own doctor because of diarrhœa and vomiting. By then he felt weak and was running a low-grade temperature. Apart from his skin lesions he exhibited at that time some epigastric tenderness which was not related to meals. Referred for a dermatological opinion on January 20, 1964, and immediately admitted to hospital.

Past history: General health always good. Measles, chickenpox and mumps as a child.

On examination (20.1.64): A thin, flushed youth of modest IQ. Regular dentition, long trunk and rather long and narrow fingers and toes. Fauces: posterior pharyngeal wall red and beefy looking, rather dry and slightly granular. No lesions seen in mouth. CVS: pulse 120 regular and of good volume. No cardiac enlargement or murmur. Lungs and abdomen normal. Skin: the neck, trunk, arms and legs exhibit a profuse eruption consisting of papular, flat and varioliform lesions varying in size from 2 to 5 mm (Figs 1 and 2). Some of these lesions were circular while others were irregularly shaped; they were characterized by a sharply-defined peripheral pink halo with a white atrophic or yellowish crusted centre. The scalp, palms and soles were spared though the forehead showed a solitary lesion. No adenopathy.

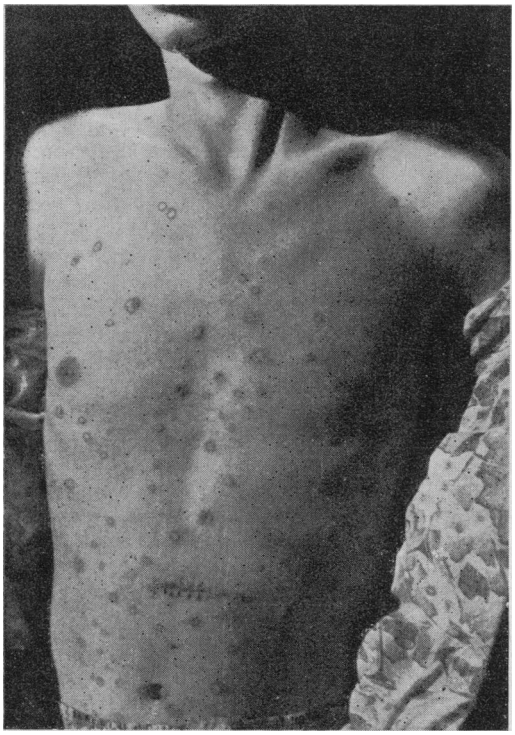

Fig 1 *Malignant atrophic papulosis: profuse and widespread eruption on trunk; the limbs were also involved*

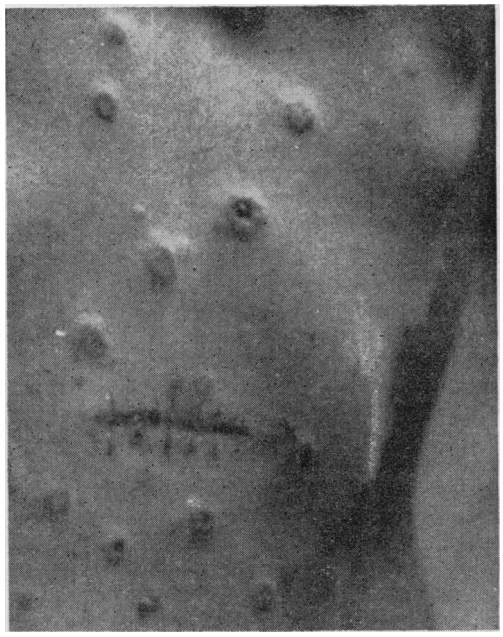

Fig 2 *Close-up around biopsy site showing lesions in all stages of development*

Investigations: Skin biopsy (Dr A R Worssam) showed an almost flat surface, with a punched out ulcer about 4 mm in diameter. Microscopic features: generalized epidermal atrophy, a homogeneous fibrinoid type of degeneration of collagen in the dermis, and a striking absence of inflammatory reaction. There was only a scanty focal lymphocytic infiltrate, often but not always surrounding arterioles, which appeared otherwise normal. No giant cell formation, skin appendages normal. There was a thrombosis of one small vein in the section, and several deeper arterioles contained no blood. Subcutaneous

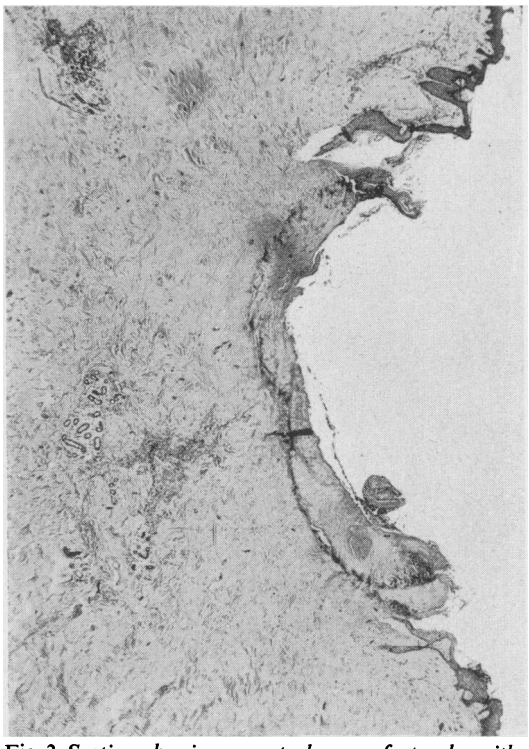

Fig 3 *Section showing a central area of atrophy with depressed ulcerated centre. There is mild lymphocytic cuffing around some of the capillaries adjacent to the atrophic area*

fat normal. In general the appearances were consistent with those of malignant atrophic papulosis described in the literature (Fig 3).

Muscle biopsy (pectoralis major) showed no abnormality, but no well-formed arteriole was present in the section. Capillaries and veins, however, appeared normal.

X-rays: Barium meal (31.1.64): stomach and duodenal cap outlined normally. The barium was followed through at one, six, twenty-four and forty-eight hours and appeared to show a fairly normal rate of progress through the bowel but the normal feathery appearance of the upper small intestine had a rather coarse appearance. Chest (23.1.64): lung fields clear.

Blood count: Hb 85% (12·4 g%). WBC 13,000 (neutros. 82%, eosinos. 1%, lymphos. 10%, monos. 7%). ESR 15 mm in 1 hour (Wintrobe). Platelets 300,000/c.mm.

Film: red cells showed some anisocytosis and were hypochromic. Serum protein: total protein 6·0, albumin 3·0, globulin 3·0 g/100 ml, A:G ratio 1:1. Electrophoresis: some increase in α_2 fraction. Very slight hypoalbuminæmia with resulting shift in A:G ratio. WR negative, precipitation test negative, no LE cells seen. Blood urea 32 mg/100 ml. Serum electrolytes: sodium 135, potassium 4·6 mEq/l. (normal). Chlorides 89 mEq/l. CO_2 combining power reduced. Fæces: occult blood test positive.

Liver function tests: bilirubin 0·8 mg/100 ml, thymol turbidity 1, thymol flocculation 1, colloidal gold 0, serum transaminase (SGPT) 14 units (normal level 5–35 units), alkaline phosphatases 11 units.

ECG 23.1.64: (1) Abnormalities suggesting posterolateral cardiac ischæmia, perhaps with posterior cardiac infarction. (2) Left ventricular preponderance.

Progress and Treatment: Gradual deterioration in general condition following admission. Anorexia and nausea with frequent vomiting of both light diet and fluids. He lost 28 lb in weight in this period. Enteric coated prednisone in dosage of 30 mg daily and antibiotics produced no change in his condition. No fresh skin lesions appeared since he was first seen.

Comment

It is thought that this syndrome has not previously been described at a meeting of the Section of Dermatology. Malignant atrophic papulosis was first recognized as an entity by Degos *et al.* in 1942 who described it as a disorder characterized by papular elements with a tendency to atrophy, and by a violent intestinal episode terminating in death. Degos (1964) in a personal communication maintains that the cutaneous lesions are so characteristic that the diagnosis can be easily made from a photograph.

Kohlmeier (1941) and Lausecker (1949) regarded the condition as intestinal thrombo-angiitis obliterans with skin lesions. 'Ulérythème porcelaine en gouttes' was the designation given by Tzanck *et al.* (1948).

The syndrome is well reviewed in the papers of Degos (1954) and those of Nomland & Layton (1960) and Naylor *et al.* (1960). Winkelmann *et al.* (1963) described two cases with cerebral involvement. From a review of the cases reported it seems that the interval between the appearance of skin lesions and the development of the fatal intestinal phase varies from three weeks to three years and is usually several months. Nonfatal cases have been described. The nosological position of the syndrome is still a matter for discussion; on pathological grounds it cannot be regarded as a manifestation of thrombo-angiitis obliterans or polyarteritis nodosa.

Postscript

Against medical advice the patient took his discharge from hospital, suffered an acute abdominal episode, and died on 28.3.64.

Summary of post-mortem report (Dr A R H Worssam): External examination: There was a sugges-

tion of Marfan's syndrome, with unusually long fingers and toes, high arches to the feet, a high roof to the mouth and prominent crowded upper front teeth. There were numerous skin lesions as described in the clinical notes. Internal examination: Brain appeared normal apart from a single area of about 1·5 in. diameter on the cortical surface of the right parietal lobe. Here there was a superficial roughened area of scarring with hyperæmia. Chest: lightly bloodstained pleural effusions on both sides of the chest. No evidence of any lesions on the visceral or parietal pleura to account for the effusions. Lungs pale with basal congestion; no naked eye lesions. Heart: pericardial sac and heart appeared normal with no myocardial scarring or abnormality of the coronary arteries. Abdomen: the skin over the abdomen showed marked post-mortem change. On opening the peritoneum, a small quantity of free bloodstained fluid was seen with coils of bowel matted together by adhesions and a plastic peritonitis, especially around the cæcum and in the lower abdomen. Only the stomach was completely free, and able to be mobilized. The stomach was empty, and showed only very small and scanty erosions, which may not be significant. The œsophagus was normal. From the duodenum to the rectum, the bowel contents were heavily bloodstained, amounting in many places to pure blood. However, an examination of the small bowel mucosa showed no localized lesion proximal to a point about 60 in. from the duodenum, where there was a crop of ulcerated lesions, similar to those seen on the skin, and associated with enlargement of the regional lymph nodes. At one point there were distinct ulcers on the periotoneal as well as the mucosal surface of the small bowel. Calcified lymph nodes about 0·5 in. in diameter were present in the mesentery close to the cæcum, but may be due to an old tuberculous infection not associated with the present condition. Further crops of mucosal ulcers were found in the terminal ileum, in the cæcum, and finally around the splenic flexure in the colon, and they were at their maximum at the last site. None was present in the rectum or sigmoid colon. All abdominal organs appeared normal except the liver, which on section showed gross fatty change amounting almost to necrosis.

Examination of the arteries and veins of both the upper and lower limbs showed no evidence of thrombosis or embolism, or of thrombophlebitis.

Cause of death: intestinal hæmorrhage.

BIBLIOGRAPHY
Church R E (1950) *Proc. R. Soc. Med.* **43**, 387
Degos R (1954) *Brit. J. Derm.* **66**, 304
Degos R, Delort J & Tricot R (1942) *Bull. Soc. franç. Derm. Syph.* **49**, 148
Howell R G (1957) *Proc. R. Soc. Med.* **50**, 1021
Kohlmeier W (1941) *Arch. Derm. Syph., Berl.* **181**, 783
Lausecker H (1949) *Acta derm.-venereol., Stockh.* **29**, 369
Naylor D, Mullins J F & Gilmore J F (1960) *Arch. Derm., Chicago* **81**, 189
Nomland R & Layton J M (1960) *Arch. Derm., Chicago* **81**, 181
Tzanck, Civatte & Sidi (1948) *Bull. Soc. franç. Derm. Syph.* **55**, 10
Winkelmann R K, Howard F M, Perry H O & Miller R H (1963) *Arch. Derm., Chicago* **87**, 54

Dr G B Dowling: This case differs from 3 others that I and others have seen abroad in that both sides of the picture are present chronically and in severe degree at the same time and the patient is very ill. The others had the same skin lesions but they were well. According to the reports the expected intestinal complication would normally be rapidly fatal.

Dr F J V Jenner: Is there anything to suggest the condition is due to drugs?

Dr I Sarkany: Dr R G Howell (*Proc. R. Soc. Med.*, 1957, **50**, 1021) showed a 14-year-old girl with the label of 'Macular atrophy of the skin' and suggested that the clinical appearances were similar to those described by Degos. However, that patient was in apparently good health. Dr Wilson-Jones has reviewed this case and tells me that in his opinion the histology would just fit with the diagnosis of malignant atrophic papulosis, but that the patient continued to attend hospital for one and a half years, remained well and did not go on producing new lesions.

Dr S P Hall-Smith: Ulcerated lesions have been described by Tzanck and others. Church (1950) showed a case here under the title of 'Dermatitis nodularis necrotica' with ulcerated lesions, but there is doubt whether this was a true case of malignant atrophic papulosis.

Dr E Wilson-Jones: Histologically the skin shows a wedge-shaped area of atrophy with an almost complete cellular black-out, but there is almost no accompanying inflammatory response within or at the margins of the lesion. There is no real histological evidence that the condition is due to a simple vascular occlusion.

Hepato-cutaneous Syndrome

I Sarkany MRCP

D S, aged 39, housewife

History: In May 1961 a rash, lasting a fortnight, appeared on her anterior chest wall and subsequently spread all over the body. This was followed by jaundice lasting nearly six months and several episodes of diarrhœa which were due to ulcerative colitis.

In November 1961 she was put on prednisolone, which was continued for six months and during this time her jaundice faded. By January 1963 she had to be readmitted to another hospital with an exacerbation of her liver condition. It was then