not much below the normal skin temperature, and precipitated in the skin venules if the ambient temperature fell below 27°C or rose to the level that induced sweating. In the later stages of her illness coagulation occurred *in vitro* at 31–34°C.

Her cryoglobulin was monoclonal in type, but our investigations failed to show multiple myeloma in life. Her skin infarcts became more and more extensive and she died towards the end of 1971. Areas of myeloma were found at necropsy in her sternum and the vertebral body of L2.

REFERENCE
Miller A J & Sheppard R F
(1970) *Proceedings of the Royal Society of Medicine* 63, 887

Dr P W M Copeman: The cutaneous signs in a patient with cryoprecipitable protein disease result from deposition of protein in the superficially placed skin venules. Venular lesions could be seen readily in Dr Sanderson's patient where the adjacent arterioles in the same neurovascular bundle were spared the blockage and angiitis (Copeman & Ryan 1971). The subpapillary venular plexus is horizontally disposed beneath the epidermis and the vessels are therefore vulnerable throughout their length to atmospheric chilling. On cooling the viscosity of blood increases, which makes it flow more slowly, and this lowered shear rate of whole blood will in itself alter the flow properties and make it yet more viscous – two mutually interacting processes. Blood stops flowing and clots with fibrin and platelets. The precise mechanism of the subsequent and rapidly developing angiitic inflammatory response is speculative.

REFERENCE
Copeman P W M & Ryan T J
(1971) *British Journal of Dermatology* 85, 205

The following cases were also presented:

Malignant Atrophic Papulosis (Degos's Syndrome)
Dr N E Jensen (for Dr R E Church)

Chronic Urticaria, Hidden Monoclonal-IgM-Protein Disease and Interstitial Pulmonary Fibrosis
Dr J J Cream (for Dr R H Meara)

(1) **Mucocutaneous Candidiasis with Thymoma**
(2) **Vasculitis with Monoclonal Gammopathy**
Dr D G C Presbury (for Dr D I Williams)

Immune Complex Cryoglobulinæmia with Cutaneous and Systemic Manifestations
Dr J J Cream (for Dr F R Bettley, Dr J D N Nabarro & Dr R Cairns)

Vasculitis
Dr J Adamson (for Dr R H Marten)

Hydroa Vacciniforme
Dr C Ramsay (for Dr P Hall-Smith)

Cheilitis Granulomatosa
Dr J M Ward
(for Dr E Cronin)

Perianal Bowen's Disease
Dr D G C Presbury
(for Dr R H Marten)

Meeting February 17 1972

Cases

'Giant' Orf of Finger in a Patient with a Lymphoma

John Savage MD
(Royal Infirmary, Doncaster)
and M M Black MD MRCP
(St John's Hospital for Diseases of the Skin, Lisle Street, London WC2)

Man aged 65

History: In September 1968 the patient presented with a tumour in the postnasal space involving base of skull and peritonsillar tissues. A biopsy showed infiltrating lymphocytic lymphoma. General examination revealed axillary lymphadenopathy and hepatosplenomegaly. The primary tumour was treated with radiotherapy and a short course of prednisone combined with immunosuppressive agents (cyclophosphamide, methotrexate and vincristine). Throughout 1969, 1970 and the early part of 1971 intermittent courses of cyclophosphamide, methotrexate and vincristine were administered intravenously. By September 1971 the liver and spleen had further enlarged and he was treated with prednisone as well as weekly intravenous injections of cyclophosphamide (1 g) and vincristine (2 mg). In early November the patient cut his left middle finger whilst cleaning out a sheep's head. The wound failed to heal and on December 17 the patient was referred to one of us (J S) because a granulomatous lesion had developed at the site of injury.

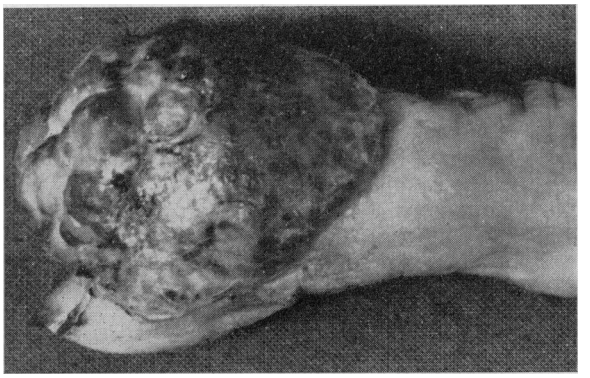

Fig 1 *Large orf lesion on dorsum of left middle finger*

On examination: Typical lesions of orf on dorsum of left middle finger and right thumb.

The lesion on the left middle finger gradually enlarged and by 12.1.72 was approximately 5 cm long and 3 cm in depth (Fig 1). After consultation with an orthopædic surgeon the left middle finger was amputated. The patient's general condition remained good.

Relevant investigations: WBC 9000/mm^3 (differential: 3500 polymorphs/mm^3). Serum immunoglobulins: IgA 68 mg/100 ml (modest reduction); IgG 750 mg/100 ml (low normal); IgM 285 mg/100 ml (modest increase). Electron microscopy (Public Health Virology Laboratory, Sheffield): orf virus present in finger tissue.

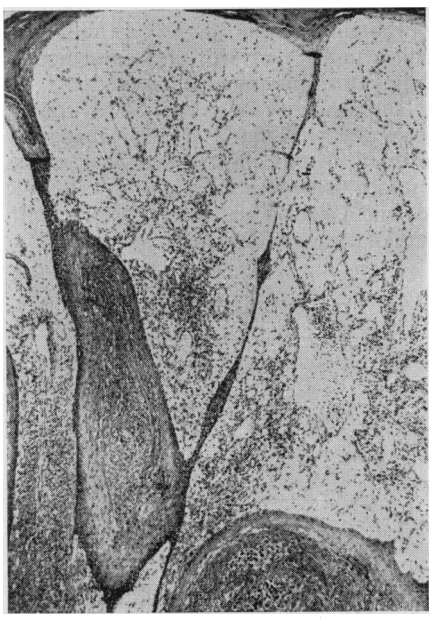

Fig 2 *Finger-like projections of epidermis penetrate the dermis. The proliferation of small blood vessels resembles that seen in pyogenic granuloma. H& E. ×13*

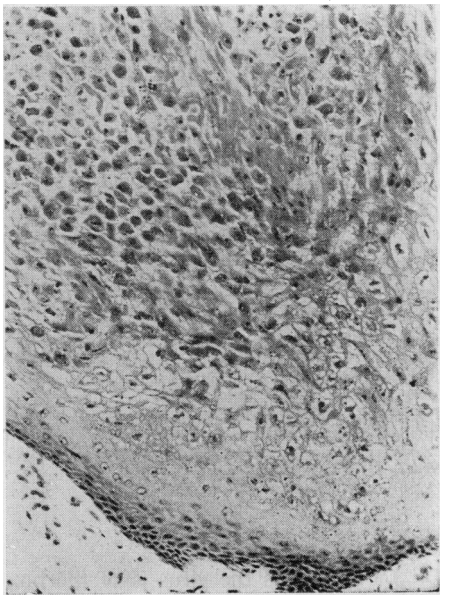

Fig 3 *Reticulate degeneration and vacuolization of upper epidermal cells, with many intracytoplasmic eosinophilic inclusion bodies. H & E. ×53*

Histopathology: Throughout the tumour finger-like projections of epidermis (Fig 2) penetrated the full thickness of the dermis. A considerable proliferation of small blood vessels was a prominent feature, which in places was similar to that seen in pyogenic granuloma (Fig 2). Vacuolization of upper epidermal cells was prominent with the formation of numerous intracytoplasmic eosinophilic inclusion bodies (Fig 3).

Comment

'Opportunist' infections from the herpes group of viruses, cytomegalovirus and wart virus are not uncommonly seen in patients treated with immunosuppressive drugs or steroids. Rimbaud *et al.* (1969) described widespread molluscum contagiosum in a man with chronic lymphatic leukæmia, and recently Ganpule & Garretts (1971) noted this association in a man with sarcoidosis. Ours is the first recorded case of an 'opportunist' viral infection due to the ecthyma contagiosum (orf) virus, occurring in a patient with lymphocytic lymphoma treated with steroids and immunosuppressive drugs.

Human orf is a benign self-limiting condition, the lesion(s) usually healing spontaneously within 36 days (Leavell *et al.* 1968). Kewish (1951) noted widespread metastatic orf lesions in a previously healthy woman, but these gradually resolved. Our patient did not develop metastatic lesions, but the typical orf on the finger continued to enlarge for two months until the finger was amputated. The

histological findings resembled those seen in the papillomatous stage of orf (Leavell *et al.* 1968). Wheeler & Cawley (1956) emphasized that the histology of orf could be similar to that of pyogenic granuloma, as in our patient. By electronmicroscopy a clinical diagnosis of orf may be confirmed within three hours because orf virus is particularly plentiful in scrapings obtained from the crusted surface of the lesion (Nagington 1964).

REFERENCES
Ganpule M & Garretts M
(1971) *British Journal of Dermatology* **85**, 587
Kewish O K (1951) *British Medical Journal* i, 356
Leavell U W jr, McNamara M J, Muelling R, Talbert W M, Rucker R C & Dalton A J (1968) *Journal of the American Medical Association* **204**, 657
Nagington J (1964) *British Medical Journal* iv, 1499
Rimbaud P, Izarn P, Meynadier J & Ravoire G (1969) *Bulletin de la Société franaçise de dermatologie et de syphiligraphie* **76**, 575
Wheeler C E & Cawley E P
(1956) *American Journal of Pathology* **32**, 535

Dr J Pegum: As the virus is of the pox group, perhaps the condition can be treated with methisazone (Marboran) which has been shown to be effective in smallpox and vaccinia infections.

Dr F A Ive: I have used 20% idoxuridine in DMSO on 4 cases of orf and formed the impression that it speeded recovery. I intend using the same treatment on infected sheep.

Dr H R Vickers: This case illustrates well the problems we are seeing increasingly of the remarkable way in which immunosuppressive drugs may modify the clinical picture of dermatoses caused by infection.

Dr A Aitken Ross: Having, in years gone by, practised dermatology in both the south-east and north-east of Scotland, which are sheep-farming areas, I was most interested to hear this report on a case of orf. I must say that from my experience of the condition the clinical appearance of the amputated finger was less like the usual flat pustule of orf and showed more of a hypertrophic granuloma. If the virus origin of the disorder were fully proven, then that is that, but I should have thought that the earlier differential diagnosis would also have embraced strawberry foot rot which is another disease carried by sheep and possibly transmissible to man. This latter is in fact a mycosis (*Dermatophilus congolensis*) and not of viral origin.

Dr J Savage: When first seen the lesions were typical of orf, presenting with flat pustules. The massive increase in size and granulomatous appearance followed. The orf virus was demonstrated in Sheffield and in London.

Anderson-Fabry Dyslipidosis

M L Johnson MRCP
(Redhill General Hospital, Redhill, Surrey)

Mr S M, aged 22

History: At the age of 11 he developed a symptomless rash on his lower trunk and buttocks and shooting pains in his feet. From the age of 13 he had shooting pains in his fingers which left his fingers feeling stiff; these pains could be provoked by raising his hands above his head. He also complained of generalized weakness especially when tired or in hot weather. The pains in the feet ceased at the age of 21 but those in the hands persisted.

Family history: The family history is summarized in Fig 1 and Table 1.

On examination: Skin: 2–3 mm angiokeratomata on genitalia and innumerable smaller similar lesions of a dark red-purple colour on the buttocks, thighs, lumbar region and lower abdomen. Fewer and smaller lesions were present all over the body including the lips and there were pinpoint angiomata on the palms. No lesions were present within the mouth but small telangiectases were seen on the face and shoulders.

Eyes (Mr F J Curtis): Diffuse fine opacities present in cornea giving the appearance of wavy marks like a sandy beach. Conjunctival vessels were normal but the retinal vessels were tortuous.

General examination was within normal limits.

Investigations: The following were normal: full blood count, serum proteins and electrophoresis, serum lipoproteins, serum electrolytes and urea, liver function tests, X-rays of chest and hands, electrocardiogram, creatinine clearance.

Urine: Doubly refractile material was present in the urinary deposit. Thin-layer chromatography of lipid extracts of this contained a gross excess of a substance with the mobility of ceramide trihexoside and a slight excess of a substance with the mobility of ceramide dihexoside.

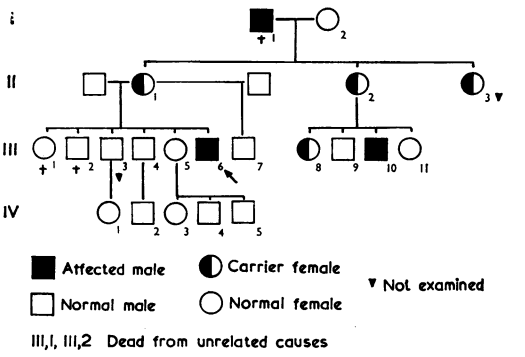

Fig 1 *Kindred of patient with Anderson-Fabry dyslipidosis*