

SOCIETY PROCEEDINGS

SWEDISH DERMATOLOGICAL SOCIETY

Meeting at Karolinska Sjukhuset, Stockholm, April 21, 1972

Generalized Plane xanthomas and Monoclonal IgG

Presented by Hans Hammar

This 62-year-old woman with two children was healthy until 1954, when she developed arthralgia in many joints, both large and small. She was treated in hospital for incipient rheumatoid arthritis. The following year xanthelasmas and plane xanthomas appeared on the trunk and subsequently increased in size and distribution. In 1966 a monoclonal IgG was found, and since that time purpuric areas have occurred especially within the plane xanthomas, leaving them increasingly pigmented.

The woman has now only small areas of normal skin surrounded by the extensive plane xanthomas. Papular xanthomas are present on the arms. Excessive xanthelasmas and folded nasal xanthomas which are markedly pigmented have developed on the face (Fig. 1).

Laboratory studies. Since 1956 the ESR has been 60-120 mm/hour; since 1964 hypochromic anemia, thrombocytopenia (ca 108 000), hyperfibrinogenemia (0.6 g/100 ml), hypergammaglobulinemia (2.6 g/100 ml) with a monoclonal band of IgG and cryoglobulinemia have been found.

Blood lipids were normal in the earlier stage of the disease and sometimes even after 1961 when the cholesterol concentration increased (450 mg/100 ml) and after 1964 when hypertriglyceridemia was found (2.8 mM). At present these values are slightly elevated but the plasma lipoproteins are within normal limits. Since 1966 repeated investigations have not revealed any internal malignancy; a lymphoma or myeloma was especially searched for.

Histological examinations have shown in the corium confluent groups of xanthoma cells which

localize perivascularly and periappendicularly together with hemosiderosis of various degrees.

Treatment. None given.

Comment. In the literature (2) references are made to dysglobulinemias associated with hyperlipoproteinemia and plane xanthomas. Many of these patients have a primary tumour or myeloma but in some cases a benign gammopathy is encountered. It cannot be decided at present to which group our patient belongs. A similar case has been reported since this meeting by Kodama et al. (1).

REFERENCES

1. Kodama, H., Nakagawa, S. & Tanioku, K.: Plane xanthomatosis with antilipoprotein autoantibody. *Arch Derm (Chicago)* 105: 722, 1972.
2. WHO Memorandum: Classification of hyperlipidemias and hyperlipoproteinemias. *Circulation* 45: 501, 1972.

Scleredema

Presented by Hans Hammar & Nils Thyresson

This 38-year-old woman had had a previous history of toxicoderma due to sulfonamides and periods of urticaria of obscure etiology. On March 3rd 1971 she became acutely ill with swelling and rigidity of the right half of the face. She had a slight fever and signs of common cold and was therefore treated with a nasal decongestant and penicillin. Two weeks later parotitis was suggested but could not be proved and the fever continued. Vertigo and faintness prompted admission to hospital, where she was treated during April and May 1971. The symptoms faded away but the facial edema remained the same. In November 1971 her face was round and swollen with varying amounts of non-pitting edema

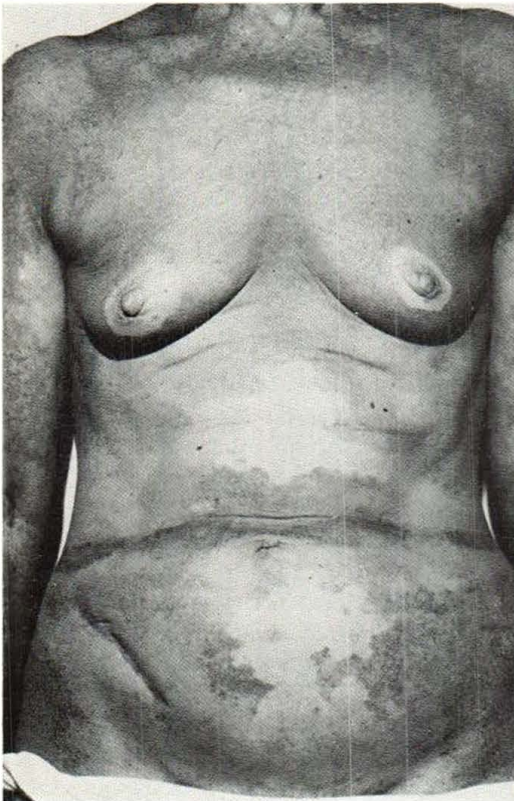
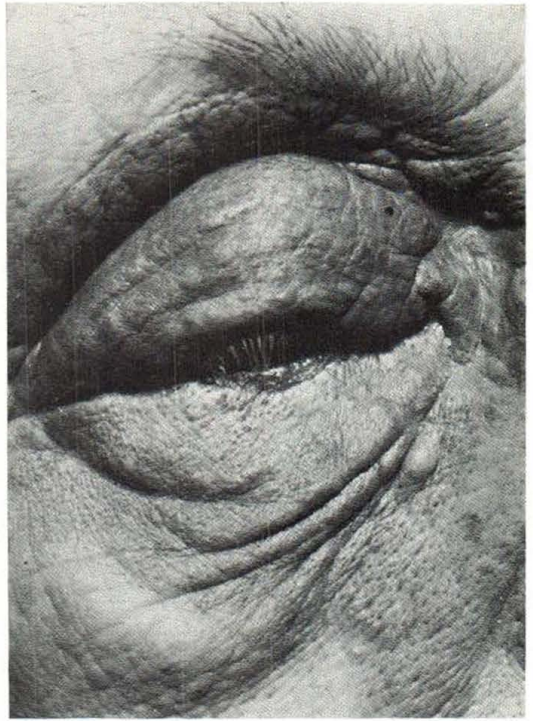


Fig. 1. Generalized plane xanthomas.



Fig. 2. Scleredema Buschke.

(Fig. 2). The lips and eyelids were not involved. She had slight edema of the arms and on proximal phalanges. The cheeks were marked with scattered telangiectases.

Laboratory studies. Analyses of the blood and urine, as well as tests of the thyroid function, gave normal results. ESR was 20 mm/hour. Liver tests revealed signs of bile stasis, due to gallstones. Cholecystectomy was performed later, after which the liver function and a liver biopsy were normal. Immunoglobulin, Coombs' direct test and serologic tests for herpes simplex, parotitis, rubella and cytomegalic inclusion disease as well as antistreptolysin and antistaphylococcus titres were all normal. PPD was positive.

Serum electrolytes, including manganese and iron were normal. The antinuclear factor was negative in April but a low titre, 1/5, was found in November.

Renal elimination was normal for mucopolysaccharides, creatine, creatinine and amino acids including hydroxyproline. Serum creatine phosphokinase was normal. ECG was normal. EMG showed slight signs of myopathy in the orbicular muscle of the eye and in the deltoid and pectoralis major muscles. X-ray examination of the facial bones, esophagus and lungs, and sialography on the left side showed nothing abnormal. Osteopoikilosis was disclosed in the skeleton of arms, legs and pelvis.

Biopsies from affected skin revealed normal

epidermis together with moderate dermal edema with small perivascular cell infiltrates, mostly lymphocytes. Staining with Alcian-blue (pH 4) showed small amounts of hyaluronic acid in the dermis. Mucicarmine-positive material was also found. The findings with these stains favour the proposed diagnosis. In some of the biopsies there was a more dense inflammatory cell infiltrate, together with many mast cells. No muscle biopsies were taken.

DISCUSSION

N. Thyresson: This case is confusing. Scleredema is indicated by the case history, yet all signs and findings fail to fit, e.g. the edema on the fingers and the variation in histo-pathology. It can be noted that the patient had osteopoikilosis as well as a son of hers. There were, however, no signs of dermatofibrosis lenticularis disseminata as in the Buschke-Ollendorf syndrome.

E. Neumann: I have seen a similar case and I am convinced that the diagnosis is scleredema of Buschke. In my case treatment with hyaluronidase was given with an excellent result.

B. Lagerholm: There were four biopsies, two were taken in April 1971 the other two in November 1971. The first two biopsies showed a considerable amount of inflammatory infiltrate, which was much less in the material obtained in November. However, in one of the earlier biopsies material stained by Alcian-blue (pH 4) and mucicarmine were found. This could be seen also in one of the later biopsies. The edematous stage in scleroderma and scleredema is, however, very difficult to differentiate histo-pathologically.

Addendum. In November 1972 the patient shows the same clinical picture as described above. Hyaluronidase has been given i.m. in a total dose of 18 000 I.U. without any clinical improvement apart from some alleviation of the pain in the facial lesion. Corticosteroids have been tried for about 2 months without effect.

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