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


Sclerodema

Presented by Hans Hammar & Nils Thyresson

This 38-year-old woman had had a previous history of toxicodermia 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.