between all doctors involved in the care of servicemen. But for good fortune, a diagnosis of insulin-dependent diabetes in this case would have ended in tragedy.

References

Primary facial lymphoedema with xanthomas

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We describe a patient who developed primary lymphoedema of the head which was associated with xanthomas. Although xanthomas are a recognized cutaneous complication of primary lymphoedema, only a few cases have been recorded and all of these have been normolipaemic. Our patient differed in having a polygenic hypercholesterolaemia, a condition not usually associated with extensive xanthomas. We believe that the chronic lymphoedema has predisposed to the deposition of lipid within the skin, giving rise to a clinically unusual pattern of xanthomas.

Case report
A 45-year-old clerk presented in 1985 with a 4-year history of facial swelling. The initial episode affected her left cheek and the swelling lasted for a week. Two weeks later she developed further swelling of the cheek and this was accompanied by periorbital oedema and swelling of the forehead. Subsequent episodes have affected the scalp and ears. Initially the swellings cleared but eventually the affected areas became persistently swollen, although the swelling fluctuates from day to day. Since 1981 she has also noticed increasing hoarseness of her voice. There was no past medical or family history of illness.

Between 1981 and 1985 the patient had been extensively investigated with the following normal or negative results: urine analysis for blood, 24-hour urine excretion of protein, full blood count, urea and electrolytes, creatinine clearance, serum C1 esterase inhibitor, serum thyroxine and serum protein electrophoresis.

Examination showed bilateral periorbital oedema (Figure 1), brawny induration of the cheeks and swelling of both ears (Figure 2). There were xanthomas on the concha of the ear (Figures 2 and 3) and in the preauricular region (Figure 2). There was no cervical lymphadenopathy. Indirect laryngoscopy showed polypoidal thickening of the vocal cords. General examination was normal.

A fasting lipid profile showed the following results: cholesterol 10.8 mmol/l (3.0-5.6 mmol/l), triglyceride 1.6 mmol/l (0.2-1.6 mmol/l). Lipoprotein electrophoresis showed an increased \( \beta \) band, a pattern consistent with a Fredrickson type IIa hyperlipidaemia. Lipid studies were performed on the patient's family and were normal.

Discussion
Cutaneous complications of primary lymphoedema include the appearance of warty hyperkeratosis and, rarely, the development of malignant angioendothelioma, a tumour which may arise in limbs affected by either primary or secondary lymphoedema. Xanthomas in association with primary lymphoedema are less well recognized and relatively few cases have been reported. In the absence of underlying disorders such as local malignant disease, previous surgery or irradiation to regional lymph nodes, our patient's lymphoedema was considered to be primary. Occasionally rosacea may be complicated by persistent facial lymphoedema and the presence of the cutaneous features of this condition...
involved sites are the genitalia and arms. Lymphoedema of the face is much rarer but is often associated with lymphoedema elsewhere.

To our knowledge, abnormalities of the vocal cords have not been recognized in association with lymphoedema of the head and neck. Indirect laryngoscopy in our patient showed polypoidal thickening of the vocal cords. The temporal relationship between the onset of the lymphoedema and the hoarseness suggests that the two conditions are related, but we accept that such degenerative changes in the vocal cords are not uncommon in middle-age and may merely be coincidental.

Previous reports of xanthomas associated with lymphoedema have described papular xanthomas arising on the affected legs of either children or adolescents. These patients were all normolipaemic, but our patient's lipid studies showed findings consistent with the diagnosis of a Fredrickson's type IIa hyperlipoproteinaemia. Negative family studies suggested that the patient had a polygenic hypercholesterolaemia rather than a familial hypercholesterolaemia (LDL receptor deficiency). Heterozygotes and homozygotes with the familial type of hypercholesterolaemia may have very florid xanthomas, especially of the tuberous and tendon xanthoma variety. In contrast, polygenic hypercholesterolaemia is usually associated with xanthelasma or a senile arcus. Our patient's xanthomas are therefore unusual in their extent and location.

In the light of previous reports we believe that chronic lymphoedema has predisposed to the development of these lesions. An incompetent lymphatic system may lead to the deposition of lipid-derived lipoproteins in the skin, which in association with the failure to remove tissue lipid may lead to the accumulation of large amounts of lipid within the skin. Support for this view comes from the observation that if lymphatic drainage is improved in primary lymphoedema by either compression or elevation, the xanthomas resolve.

References

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