

DERMATOLOGY AND SURGERY

R.O. PARNIS

M.B.E., M.D., F.R.C.S.

In this paper I shall describe two dermatological patients who came my way recently. The first one was a handsome, fair-haired, lusty individual in his mid-twenties who had been attending the skin clinic for years, certainly for eight years. He emitted a very disagreeable smell which came from numerous sores all over the back but mainly on the shoulders and buttocks. The story was that these ulcers healed only to break down again, they appeared on lumpy skin and they became active with no obvious cause. This man like the woman troubled with an issue of blood 'suffered many things from many physicians and had spent all that she had and was nothing the better but rather worse'. He did not improve because his complaint was acne conglobata. This is an unusually severe form of acne, very rare, the hallmarks of which are deep abscesses and draining sinuses. There are numerous comedones, cysts containing a clear viscid fluid and grouped inflammatory nodules. Pronounced scars, frequently keloidal, remain after healing. This severe disease occurs mainly in males around 16 years old and it may last even to the forties, especially on the back. The condition is extremely persistent and slow healing of one group of abscesses may accompany the progressive extension of another group of lesions. Suppurative hydradenitis and dissecting cellulitis of the scalp are sometimes associated with acne conglobata and squamous carcinoma may rarely develop in areas of scarring. The treatment which includes steroids in very severe cases is palliative and unrewarding.

This patient was referred to St. John's Hospital for Diseases of the Skin in London and after a short stay there he was sent to a plastic surgery centre where the worse affected skin over both buttocks was removed and replaced by split skin from the back of both thighs, trouble free areas.

He was also given daily injections of oestrogens and of ACTH. He has recently returned to Malta and I propose to carry on the treatment in the shoulder areas.

The second patient was a nervous young man of 25, rather thin who had yellowish patches over elbows and knees and big yellow pedunculated tumours hanging from each buttock. He had a very marked corneal arcus and his serum cholesterol level was up in the 400's. No diminished sugar tolerance was present. We are dealing here with one of the lipoidoses, conditions associated with a disturbance of lipoid metabolism. The subgroup that interests us is that of essential familial xanthomatosis which is characterised by consistent abnormalities of serum lipid concentrations, hereditary transmission to offspring and typical cutaneous manifestations. There is a disorder of cholesterol metabolism but its cause is unknown. As regards transmission, the exact mechanism of inheritance is still disputed but it appears to be either a dominant or an incomplete dominant trait. The form this patient exhibited, xanthoma tuberosum, affects young adults of either sex. The eruption consists of yellowish papules, nodules, plaques and tumours located chiefly on extensor surfaces. Lesions tend to group about the large joints, in our case the elbow and knees, and over areas of trauma and pressure such as the buttocks. Occasionally they are scattered all over the trunk. By gradual enlargement and meeting of groups of papules large pedunculated tumours may form as happened in this case over the buttocks. These tumours may show pressure atrophy of the overlying skin, telangiectasis of superficial vessels and the occasional development of stubborn painful ulcerations. The blood serum shows a marked increase in the cholesterol values as it did in our case but the figures

may sometimes reach four or five times the normal. The serum is usually clear or only slightly opalescent and the fatty acids and phospholipids are normal or nearly so. Arteriosclerotic vascular disease is common and early. Half the patients develop coronary atheroma with angina. Peripheral vascular disease is often seen. The vascular complications are the most serious cause of disability and it is said that every case of coronary artery occlusion in children and adolescents reported in the literature has been associated with xanthoma tuberosum. Actual xanthomatous nodules consisting of typical foam cells occur almost solely in the skin. Rare instances of organ involvement with epilepsy, for example, have been reported. The only exception is the vascular tree: some investigators believe that the plaques in this disease represent xanthomatosis of the arteries. Because of its potentially serious prognosis xanthoma tuberosum should be distinguished carefully from other similar conditions but this is not usually difficult.

In the case described the patient was

referred to the surgical department for removal of his buttock tumours but he was much more concerned with the flat yellow plaques, relatively inconspicuous at the elbows, and I removed a good part of these being able to do so by reason of the laxity of the skin. The wounds healed without trouble. As regards his family, his mother is alive and well aged 54 and he has a sister without xanthomas. His father died suddenly at the age of 48 while out at work and a younger brother who had lesions similar to his died suddenly at the age of 16 while out hiking. This man is engaged to be married and I am very glad I have not been asked my advice as to the wisdom of this step.

Treatment tries to deal with the underlying disease but as this is unknown we are groping in the dark. Very low fat diets are recommended, diets which are monotonous to eat, difficult to prepare and hard to pursue. In addition such diets must be constantly and strictly followed. The lumps and swellings can of course be removed for temporary benefit.