PACHYDERMOPERIOSTOSIS AND LEPROSY

Sir.

I write to propose that pachydermoperiostosis be given a recognized place in the differential diagnosis of lepromatous leprosy (LL). Apart from my references to this disease^{1,2} there has been no mention of it in leprosy literature, and it is time leprosy workers were given a description of it. In the *Textbook of Dermatology*, 4th edition, it is called 'a rare developmental defect which has been reported in many races, and occurs predominantly in males. The condition has been attributed to an autosomal dominant gene of variable expressivity'.

Physical signs of primary pachydermoperiostosis. These are largely confined to skin and bones. There is thickening of the skin of the scalp, forehead, and to a lesser extent, of the face, with a pattern of folds and fissures simulating the leonine facies of LL. There is thickening of the skin of the hands and feet, and clubbing of fingers and toes. Thickening of upper eyelids gives an expression of somnolence, and the nose may enlarge. Facial skin is shiny because of sebaceous activity, and sweating may be excessive. There have been reports of gynaecomastia and impotence. X-rays show periosteal thickening (proliferative periostitis) of limb bones. Skin and bone changes develop soon after puberty, progress for the next 5–10 years, then remain unchanged (see Table 1). Secondary pachydermoperiostosis causes no confusion with leprosy as skin changes are mild or absent, and the patient has chronic pulmonary disease.

A Case Report. An Indian male in his early 30s was referred to me at the Hospital for Tropical Diseases in 1973 with a provisional diagnosis of LL. On examination he looked older than his years. The skin of his forehead was thickened and thrown into folds, and his upper eyelids were thickened

Table 1. Similarities between pachydermoperiostosis and lepromatous leprosy

| Pachydermoperiostosis | Lepromatous leprosy |
|---|--|
| It may appear in families | It may appear in families |
| Patients look prematurely old | Patients look prematurely old |
| Forehead is furrowed | Forehead is furrowed |
| Upper eyelids thickened | Upper eyelids thickened |
| Face appears shiny | Face appears shiny |
| Nose may be enlarged | Nose may be enlarged |
| Legs and feet become thickened | Legs and feet become thickened |
| Gynaecomastia and impotence may develop | Gynaecomastia and impotence may develop |
| Hyperhidrosis common | Hyperhidrosis (compensatory) common |
| Peripheral nerves thickened | Peripheral nerves thickened in late stage |
| Periostitis of long bones | Periostitis of long bones in neglected cases |

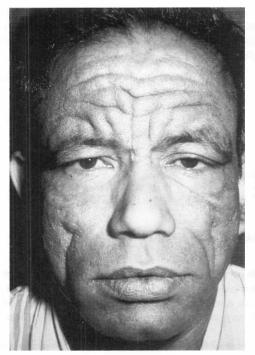


Figure 1. The patient.

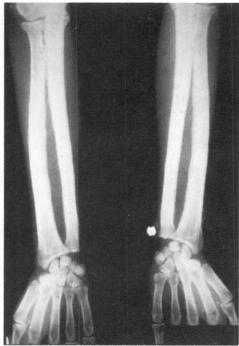


Figure 2. Proliferative periostitis of radius and ulna.

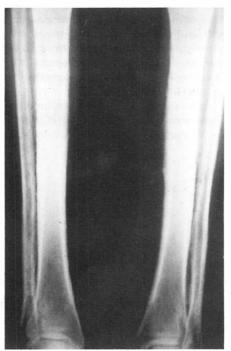


Figure 3. Proliferative periostitis of tibia and fibula.

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(Figure 1). The skin of his hands and feet was thickened and there was clubbing of fingers and toes. X-rays of bones of forearms and lower legs showed periosteal thickening (Figures 2 and 3). Palpation of his peripheral nerves revealed generalized thickening—a feature which had not previously been recorded in the literature. Skin smears were negative for AFB.

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References

¹ Jopling WH. Handbook of Leprosy. London: W Heinemann Medical Books, 1984.

² Jopling WH, Harman RRM. Leprosy. In: *Textbook of Dermatology*. Rook A, Wilkinson DS, Ebling FJG, Champion RH, Burton JL, (eds), Oxford: Blackwell Scientific Publications, 1986, 823–38.