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NEUROFIBROMATOSIS WITH UNIVERSAL PERIPHERAL NERVE INVOLVEMENT

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A 20-year-old man was referred to our department with universal nerve thickening and beading for the exclusion of Hansen's disease. On examination he had multiple soft mollusca fibrosa lesions, cafe-au-lait macules over the face and trunk along with Lisch nodules in both the eyes. There was no neurological deficit. Slit skin smear for acid fast bacilli was negative. Sural nerve biopsy showed neurofibroma lesions.

Key Word : Neurofibromatosis

Introduction

Neurofibromatosis is a heredofamilial disease characterised by cutaneous pigmentation and multiple tumours arising from the peripheral and central nervous system which are mainly hamartomas arising from the neuroectoderm.[1] Involvement of peripheral nerves by tumours is one of the usual features of Von Recklinghausen's disease.[1] In contrast to the intracranial or intraspinal tumours, involvement of major peripheral nerves does not necessarily constitute a significant clinical finding.[1] Any of the major nerves in the neck, trunk, extremities and certain cranial nerves along their peripheral course may be involved.[1] The ulnar and radial nerves are said to be the major nerves most often involved.[1]

We report a case of neurofibromotosis which had uniform and gross involvement of all the peripheral nerves.

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Case Report

A 20-year-old man was referred to the skin department for exclusion of Hansen's disease. Multiple, discrete, some hyperpigmented and some skin coloured, smooth, soft, sessile non-tender nodules with well-defined margins varying in size from 1/2 cm to $1^{1/2}$ cm were present on the face and trunk for 4 years. Multiple cafe-au-lait macules and

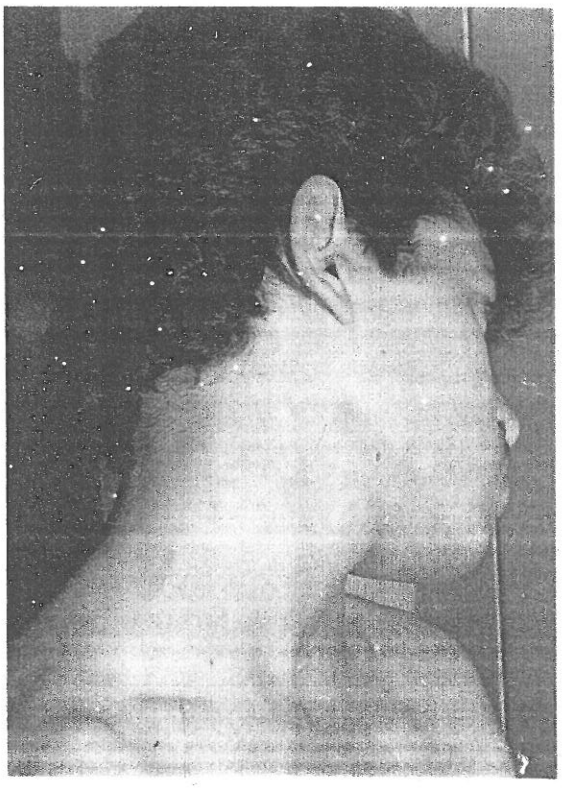

Fig. 1. Thickening and beading of the nerves

hyperpigmented macules of variable sizes were also present over the face, trunk and lower extremities. Family history of similar lesions in the mother was present. All the peripheral nerves including finer nerves like the intercostal nerves and the cutaneous nerves of the forearm and thigh were involved. The nerves showed marked thickening and beading along their course (Fig. 1). There was no neurological deficit inspite of such a marked thickening of all the peripheral cutaneous nerves.

Sural nerve biopsy revealed a neurofibroma. It was negative for any amyloid deposits. Slit skin smears were negative for AFB. Routine investigations and radiographs were unremarkable. However CSF analysis revealed marked elevation of proteins and Pandy's test was positive. Sugar, chloride and cell count were within normal limits. Ultrasound examination of the abdomen revealed retroperitoneal, intermuscular and subcutaneous tumours. Slit lamp examination confirmed the presence of Lisch nodules in both eyes. Audiometry was unremarkable.

Comments

Such universal nerve involvement cases of neurofibromatosis is rare.[1] could find only one report with simil peripheral nerve involvement reported an Indian journal.[2] The literature on th incidence of peripheral nerve involvemen is also very scarce. This is mainly becaus much importance is not given to the site and degree of involvement of the peripheral nerves as the tumours frequently are asymptomatic.

In this case the nerve involvement was so universal and gross that it mimicked nerve involvemment in Hansen's disease except for the absence of neurologic deficit.

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