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Clinical Images

Plexiform neurofibromatosis type 1

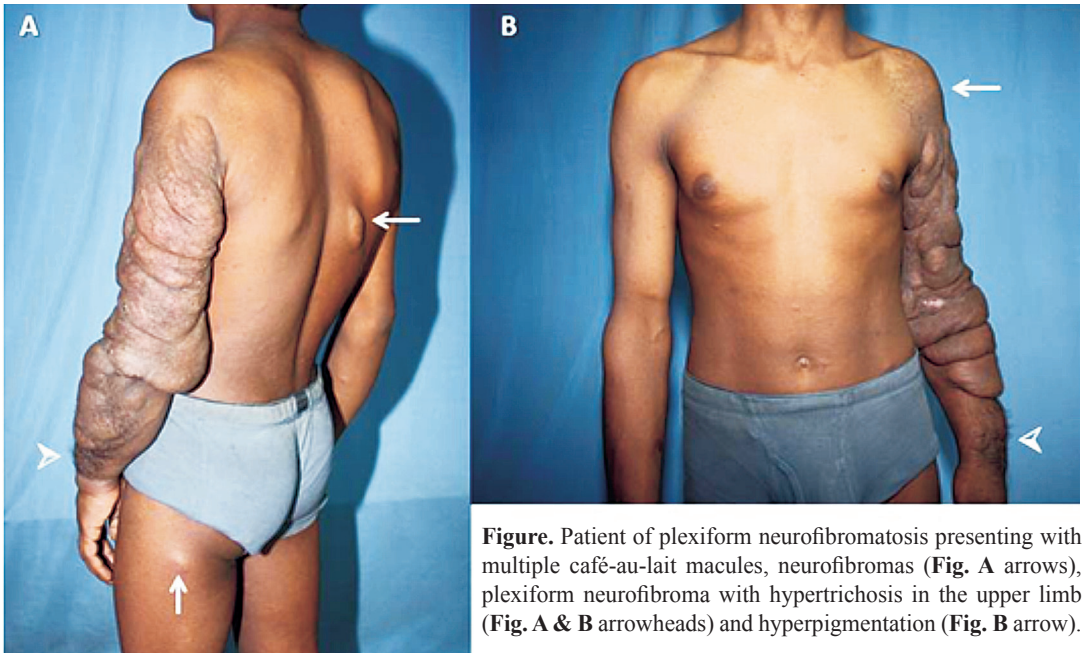

Figure. Patient of plexiform neurofibromatosis presenting with multiple café-au-lait macules, neurofibromas (**Fig. A** arrows), plexiform neurofibroma with hypertrichosis in the upper limb (**Fig. A & B** arrowheads) and hyperpigmentation (**Fig. B** arrow).

A 16 yr old boy presented to the department of Medicine, All India Institute of Medical Sciences (AIIMS), New Delhi, India, in 2013 with primary cosmetic concern about swelling in the left upper limb which was progressively increasing over the last few years. A rigorous clinical work-up revealed presence of several café-au-lait macules (CALM) (>5) along with multiple neurofibromas (Fig. A arrows), plexiform neurofibroma of the left upper limb, hypertrichosis (Fig. A & B arrowheads) and hyperpigmentation (Fig. B arrow). Magnetic resonance imaging (MRI) of the adrenals, brain and spine was non-contributory. There was no cognitive impairment, signs of skeletal deformities, ocular and acoustic abnormalities or any family history of neurofibromatosis. A diagnosis of plexiform neurofibromatosis type 1 was made which is an autosomal dominant disorder. Plexiform neurofibroma has been classically described as a "bag of worms". Amongst the complications, malignant transformation although rare, has been reported and due to progressive increase in size of the plexiform neurofibroma, disfigurement and bony erosions can occur. No definitive treatment was offered at this time. However, a regular follow up was advised keeping in mind the risk of future complications such as hypertension, *etc*.

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