Diffuse plexiform neurofibroma

S. Sujith Kumar · D. Muthiah Selva Kumar · R. Thirunavukarasu

Received: 13 November 2009 / Accepted: 31 January 2010
© Association of Surgeons of India 2010

Case report

A 15-year-old girl presented with a swelling over right thigh for a period of 3 years. She has multiple cafe – au – lait patches over the back ranging from 1.5 to 2 cm in size. On examination of right thigh, there is a large, soft, compressible, loosely hanging, hypertrichotic and hyperpigmented plaque measuring around 25 × 18 cm. It extends over the upper and middle part of the thigh in the anterior aspect (Fig. 1). On examination of the back, multiple, small, molluscum tumours were distributed and axillary freckling were present. Routine investigations were normal.

Patient was diagnosed to have neurofibromatosis Type I with large diffuse plexiform neurofibroma of right thigh. Patient underwent excision of diffuse plexiform neurofibroma (Fig. 2) and the wound was closed partially by primary closure and remaining by split skin grafting (Fig. 3). Skin graft taken well and wound healed completely (Fig. 4).

Discussion

Diffuse plexiform neurofibroma also known as elephantiasis neurofibromatosa [1], has an overgrowth of epidermal and subcutaneous tissue associated with a wrinkled and pendulous appearance. Plexiform neurofibroma occurs only

Fig. 1 Preoperative picture of diffuse plexiform neurofibroma of right thigh

Fig. 2 Excised specimen of diffuse plexiform neurofibroma

S. S. Kumar · D. M. Kumar · R. Thirunavukarasu
Department of General Surgery,
Vinayaga Missions Medical College and Hospital,
Karaikal - 609602, Pondicherry U.T.

S. S. Kumar (✉)
E-mail: docsuji1977@yahoo.co.in

Fig. 3 Immediate postoperative picture showing partial primary closure with split skin grafting

Fig. 4 Late postoperative picture

in 5% of patients with neurofibromatosis Type I [2]. Most common sites for plexiform neurofibroma are orbital and periorbital regions [3]. Its presence over the lower extremity (a rare site) is an interesting feature in our case.

Neurofibroma Type I is a common, autosomal-dominant disorder, with an incidence of about 1 in 2,500–3,300. It accounts for up to 90% of the cases of neurofibroma. The cardinal clinical features are multiple neurofibromas and pigmentary changes. These include café-au-lait spots, freckles, and hyperpigmentation overlying plexiform neuro-

fibromas. A century ago von Recklinghausen [4], described neurofibromatosis in his patients Marie Kientz and Michael Bar. Broadly, it has been divided into neurofibromatosis Type I and Type II. Four varieties of neurofibromas have been documented, cutaneous neurofibroma, subcutaneous neurofibroma, nodular plexiform neurofibroma and diffuse plexiform neurofibroma.

Plexiform neurofibroma is considered to be a hamartoma rather than a typical tumor [5]. Plexiform neurofibromas are diffuse, elongated fibromas coursing along the nerves. These lesions frequently involve the trigeminal or upper cervical nerves. Plexiform neurofibromas often appear within the first 2 years of life. Complications of plexiform neurofibroma are malignant change, bleeding from trauma, neurological deficits, limited limb movements, and psychological disturbance because of abnormal anatomy. The complications can be treated by surgical intervention. Excessive and dangerous bleeding is one of the complications while doing surgery for this condition.

Our case presented with neurofibromatosis Type I with large, diffuse, plexiform neurofibroma of right thigh. The presence of hyperpigmentation and hypertrichosis simulated giant hairy naevus. Our patient underwent excision with partial primary closure and split skin grafting. Histopathological examination confirms the diagnosis of plexiform neurofibroma.

References

1. Harper JI (1998) Genetics and genodermatoses. In: *Text book of Dermatology 6th edition*, Champion RH, Burton JL, Burns DA, Breathnach SM (Eds.) Rook/Wilkinson/Ebling. Oxford: Blackwell Science pp 378–384
2. Pivnick EK, Riccardi VM (1999) The Neurofibromatoses. In: *Fitzpatrick's Dermatology in General Medicine*, Freedberg IM, Eisen AZ, Wolff K, Austen KF, Goldsmith LA, Kartz SI, Fitzpatrick TB (Eds.). New York: McGraw Hill pp 2152–2158
3. Riccardi VM (1981) Von Recklinghausen neurofibromatosis. *New Engl J Med* 305:1617–1627
4. Von Recklinghausen FD (1882) Ueber die in multiplen Fibrome der Haut und ihre Beziehung zu den multiplen Neuromen. Berlin: Hirschwald
5. Sengupta SP (1996) Tumours and cysts. In: *Long and short cases in surgery* 1st edition, Calcutta: New Centre Book Agency Publications pp 39–75