Spectrum of Surgical management of giant neurofibroma in soft tissue: a single-center prospective analysis in Malwa region: central India

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Abstract

Neurofibroma, a common benign soft tissue tumor may continue to grow and may attain giant size. We prospectively analyzed surgical management of 10 patients with giant neurofibroma in our medical college from Oct- 2014 to Oct- 2015. The tumors were located in the head and neck (n = 2), limbs (n = 5), and retroperitoneal (n = 2), perineal region (n=1). According to the location and extent of the lesion, surgical management was performed to resect the tumor and wounds were closed either by skin flap or by skin graft. One patient of giant tumor in the esophagus was subjected to wide excision and primary repair. Post surgery the appearance of the patients and also the function of the limbs improved remarkably. In cases of giant neurofibromas, surgical treatment effectively reduces the tumor burden, rehabilitates the appearance, function and so improves the quality of life. Regular follow up and self examination of operated limb helps to diagnose early recurrence.

Keywords: Neurofibroma, Giant neurofibroma, Benign, operation, wide excision

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Introduction

Neurofibroma, a common benign tumor, may occurs in various sites of body. Most of the neurofibromas are single[1]. Syndromes associated with Multiple neurofibromas are called neurofibromatosis. Neurofibromatosis are three types, type 1 neurofibromatosis (NF1), type 2 neurofibromatosis (NF2) and schwannomatosis [1,2]. NF1 often involve soft tissue. The tumor may continues to expand so it often appears to be giant, which is a big problems in respect of cosmectic and pressure necrosis. Ulcer and repeated bleeding of the tumor may distress patients and affect quality of life. Unfortunately, although we have known something about its pathogenesis [3-6], surgical management is still the most important method to control or even cure this tumor [7,8]. Giant neurofibroma, is always a challenge due to the brisk bleeding in the operation and the difficulty of reconstruction. In this study, we made a prospective analysis of giant neurofibroma who underwent surgical management from Oct -2014 to Oct- 2015.

Methods and material

This research was conducted in our MEDICAL COLLEGE. Informed consent was taken from patients. Two patients out of ten patients meet the diagnostic criteria for neurofibromatosis type 1 [1]. From oct. 2014 to oct. 2015, 10patients (7males and 3 females) of giant neurofibroma were taken into the study. Ages of patients at admission ranged from 8 years to 60 years old. The tumors were located in the head and neck (n = 2), limbs (n = 5), and retroperitoneal (n = 2), perinealregion (1).Two patient had tumors at multiple sites.8 patients had solitary lesion, and2patients were diagnosed as NF1. All patients underwent preoperative profile including blood reports, and relevant fitness, and radiological evaluation. Tumor extension and relevant anatomy around it were displayed. Neurofibroma of viscera need proper surgical work up and nutritional build up prior of surgery.

Surgical management was based on the information from above examinations and work up. For the tumor in the trunk or limbs, the aim of surgical management was to reduce the tumor burden with functional limb. We removed the tumor completely and reconstruction done by flap or skin graft. For the tumor in the head and neck, the appearance was very concerned so we tried to make a complete resection of the tumor as far as possible with proper reconstruction. After operation, vitals were monitored until the patients recovered well.

Result

All of 10 patients underwent successful surgical management. Partial resection of the tumor was performed in 6 cases, and complete resection in 4 cases. The wounds were closed by the flap or skin graft. Among them, one female with a neurofibroma in the perineal region was treated by wide excision and

rotation flap, one adult male with esophageal neurofibroma managed with esophagotomy and wide excision. Out of these ten details of 5 cases.

S.No.	A/S	Chief complaints	Procedure	Histopath.
Case 1	50/m	Painless swelling in neck since 6 months	Esophagotomy and Wide excision of tumor with esophageal closure	Neurofibroma (plexiform)
Case 2	34/m	Rt. Upper limb swelling since 3 years	Wide excision with grafting	neurofibroma
Case 3	60/m	Swelling over rt elbow joint since 6 months	Wide excision with split thickness skin graft	neurofibroma

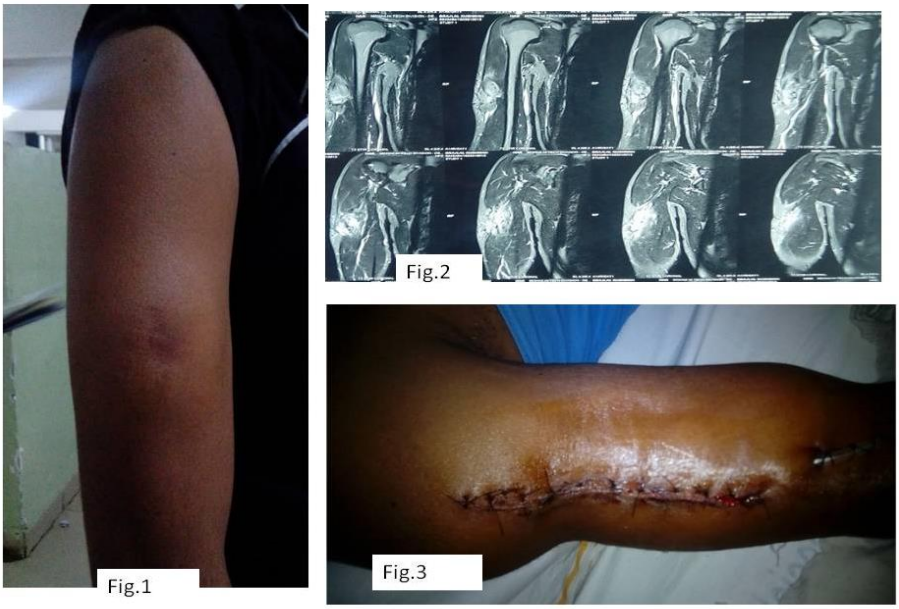

Fig. 1: esophageal neurofibroma managed successfully with esophagotomy and wide excision.

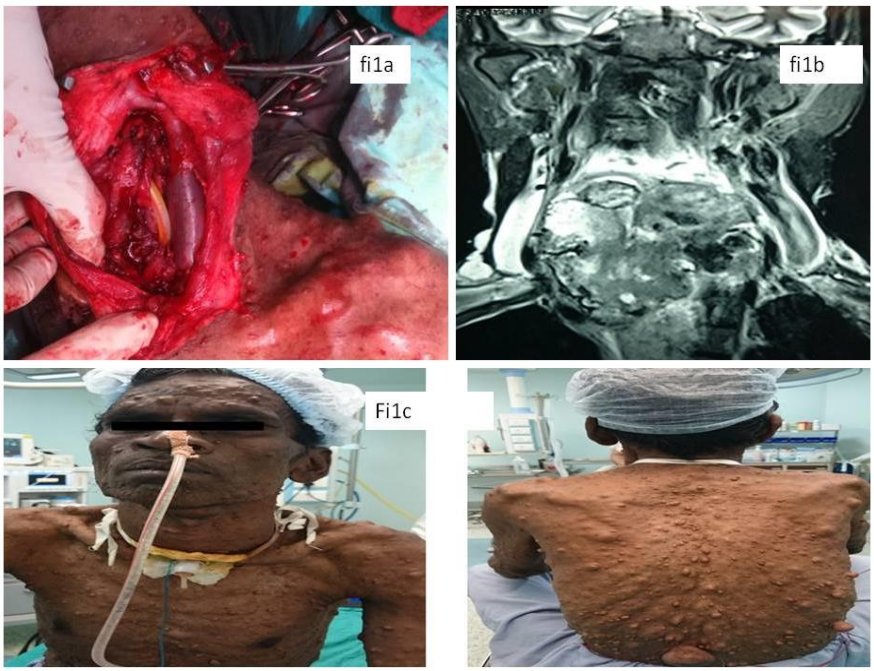

Fig. 2: neurofibroma of arm (at level of upper humerus) managed successfully with wide excision

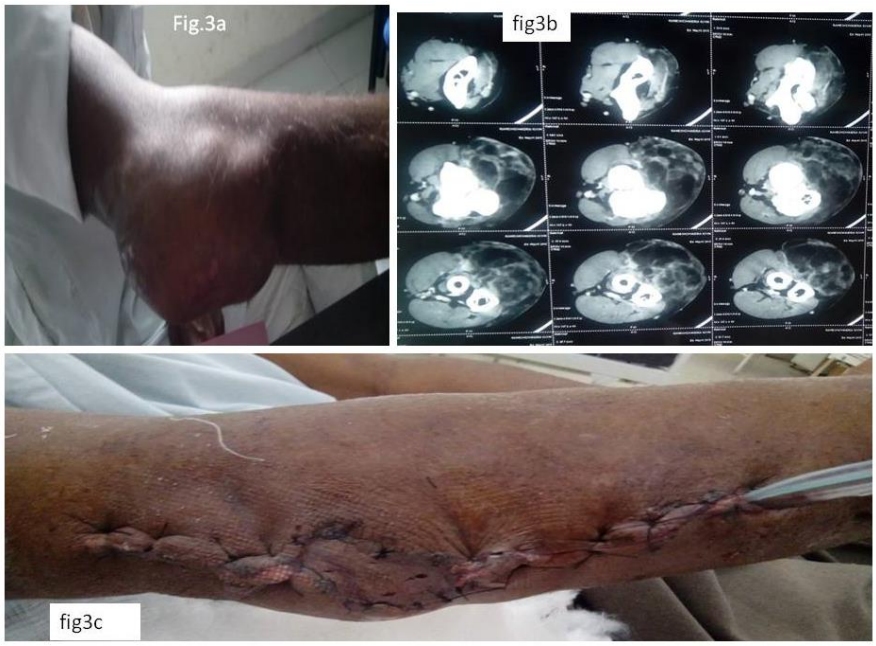

Fig. 3: Giant neurofibroma of elbow joint managed successfully

Discussion

Most of the neurofibromas are sporadic, however some of them transmitted as autosomal dominant disorder. Abnormal differentiation of neural crest cells responsible for genetically transmitted disease. NF1(17q11.2), NF2(22q11.2q12) and schwannomatosis (SMAR-CB1) are classical three types of neurofibromatosis, which arises due to mutation in their respective chromosome(1,2,9-12). NF1 (Von Recklinghausen disease) mostly affects skin and soft tissue, there is no significant difference in incidence between male and female population. Growth of Neurofibroma not uniform, usually present with irregular boundary and surface. Surgery is the most effective definitive treatment for neurofibroma. Adequate Knowledge of anatomy and location of lesion is important for surgical treatment.(3-8)

Esophageal growth managed with esophagotomy and wide excision with primaryclosure (case1). Perineal neurofibroma addressed to wide excision and closure with (case 4) rotation flap. Small tumorwas addressed to resection of the tumor with skin grafting (case 2, 3) fig 2, 3.Among abdominal neurofibroma, plexiform is most common type and paraspinal and spinal are common sites for abdominal neurofibroma (8).

Incidence of Gastrointestinal involvement in NF1 is reported to 10%–25% (9). Gastrointestinal neurofibromas are mostly asymptomaic(11). Patient with lump in abdomen under psoas muscle, managedwith almost complete wide excision (case5).

Pathologically Neurofibromas are classified as localized, plexiform, or diffuse. At gross inspection localized intraneural neurofibromas are well-defined fusiform or diffuse lesions that appear confined to the affected nerve (7). They appear gray to tan on cut sections withfocal areas of heterogeneity reflecting variable collagen content. Microscopically neurofibromas are composed of Schwann cells and fibroblasts in a myxoid.

Plexiform neurofibromais mostly seen in NF1 with more chance for malignant transformation to MPNST. Plexiform neurofibromas affect medium- to large-sized nerve. Nerves not loss original configuration. The term *ropelik*and bag of worm has been applied to the macroscopic appearance of plexiform neurofibromas that involve nonbranching nerves and highly branching nerves respectively (7).

The plexiform neurofibromas are majority of neurofibromas affecting the abdominal viscera and mesentery in patients with NF1. The majority of gastrointestinal neurofibromas in NF1 are focal intraneural or plexiform neurofibromas that involve the myenteric plexus. (13). In CT majority of neurofibromas are smooth, round or tubular masses that are homogeneously hypoattenuating on intravenous contrast (8,14). myxoid and mucinous stroma that can be observed microscopically within these tumors responsible for low attenuation in CT (7,15). Excessive collagen may result in focal areas of hyperattenuation. Tonsgard et al (8) reported observing intravenous contrast enhancement in 50% of their patients with abdominal or pelvic plexiform neurofibromas who had both nonenhanced and intravenous contrast-enhanced CT scans. Case 1 was unique in its site (esophagus), size (about 8 × 8 cm) with dysphagia without neurological deficit(case1)(16,17).

Case 2 was presented with a painless swelling of the left arm, on evaluation, tumour was deep seated and the X-ray showed erosion of the humerus. There were no “cafe au lait” spots, two or more neurofibromas,

Lisch nodules, axillary or inguinal freckling, sphenoid wing dysplasia or thinning of the cortex of long bone, and optic glioma. Wide excision was planned with split skin grafting. Final histopath suggestive of neurofibroma(18). Regular follow up and self examination is crucial for early recurrence of tumor.

In summary, this prospective study shows that individual surgical management of giant neurofibroma effectively reduces the tumor burden, rehabilitates the appearance and function, and so improves the quality of life. Post op physiotherapy help to early recovery(19-21).

Reference

1. Kopf BR. Neurofibromatosis. Handb Clin Neurol 2013; 111: 333-340.
2. Chen SL, Liu C, Liu B, Yi CJ, Wang ZX, Rong YB, Zhu J, Ding Y, Tian GL. Schwannomatosis: a new member of neurofibromatosis family. Chin Med J 2013; 126: 2656-2660.
3. Friedrich RE, Beer C, Glatzel M, Hagel C. Vascular endothelial growth factor, basic fibroblast growth factor and epithelial growth factor receptor in peripheral nerve sheath tumors of neurofibromatosis type 1. Anticancer Res 2015; 35: 137-144.
4. Chen Z, Liu C, Patel AJ, Liao CP, Wang Y, Le LQ. Cells of Origin in the Embryonic Nerve Roots for NF1-Associated Plexiform Neurofibroma. Cancer Cell 2014; 26: 695-706.
5. De la Croix Ndong J, Makowski AJ, Uppuganti S, Vignaux G, Ono K, Perrien DS(5), Joubert S, Baglio SR, Granchi D, Stevenson DA, Rios JJ, Nyman JS, Elefteriou F. Asfotase-α improves bone growth, mineralization and strength in mouse models of neurofibromatosis type-1. Nat Med 2014; 20: 904-910.
6. Rahrmann EP, Moriarity BS, Otto GM, Watson AL, Choi K Collins MH, Wallace M, Webber BR, Forster CL, Rizzardi AE, Schmechel SC, Ratner N, Largaespada DA. Trp53 aploinsufficiency modifies EGFR-driven peripheral nerve sheath tumori
7. ScheithauerBW, Woodruff JM, Erlandson RA. Tumors of the peripheral nervous system. Washington, DC: Armed Forces Institute of Pathology, 1999.
8. TonsgardJH, Kwak SM, Short MP, Dachman AH. CT imaging in adults with neurofibromatosis-1: frequent asymptomatic plexiform lesions. *Neurology*1998; 50: 1755–1760. CrossRef, Medline.
9. RiccardiVM. Neurofibromatosis: phenotype, natural history, and pathogenesis. 2nd ed. Baltimore, Md: Johns Hopkins University Press, 1992.
10. RiccardiVM. Neurofibromatosis: phenotype, natural history, and pathogenesis. 2nd ed. Baltimore, Md: Johns Hopkins University Press, 1992.
11. ScopusR. F. Gmyrek, R. Beer, D. N. Silvers, R. Reiffel, and M. E. Grossman, "Periungual myxoid neurofibroma," Cutis, vol. 69, no. 1, pp. 54–56, 2002. View at Google Scholar.
12. PetersenJM, Ferguson DR. Gastrointestinal neurofibromatosis. *J Clin Gastroenterol*1984; 6: 529–534. CrossRef, Medline.
13. Janes LE, Sabino J, Matthews JA, Papadimitriou JC, Strome SE, Singh DP. Surgical management of craniofacial neurofibromatosis type 1 associated tumors. J Craniofac Surg 2013; 24: 1273-1277.
14. Latham K, Buchanan EP, Suver D, Gruss JS. Neurofibromatosis of the Head and Neck: Classification and Surgical Management. Plast Reconstr Surg 2015; 135: 845-55.
15. Ledbetter DH, Rich DC, O'Connell P, Leppert M, Carey JC. Precise localization of NF1 to 17q11.2 by balanced translocation. Am J Hum Genet 1989; 44: 20-24.
16. Hulsebos TJ, Bijlsma EK, Geurts van Kessel AH, Brakenhoff RH, Westerveld A. Direct assignment of the human beta B2 and beta B3 crystallin genes to 22q11.2-q12: markers for neurofibromatosis 2. Cytogenet Cell Genet 1991; 56: 171-175.
17. Plotkin SR, Blakeley JO, Evans DG, Hanemann CO, Hulsebos TJ, Hunter-Schaedle K, Kalpana GV, Korf B, Messiaen L, Papi L, Ratner N, Sherman LS, Smith MJ, Stemmer-(17)Rachamimov AO, Vitte J, Giovannini M. Update from the 2011 International Schwannomatosis Workshop: From genetics to diagnostic criteria. Am J Med Genet A 2013; 161A: 405-416.
18. Ferner RE, Gutmann DH. Neurofibromatosis type 1 (NF1): diagnosis and management. Handb Clin Neurol 2013; 115: 939-955.
19. Lloyd SK, Evans DG. Neurofibromatosis type 2 (NF2): diagnosis and management. Handb Clin Neurol 2013; 115: 957-67.
20. Marocchio LS, Oliveira DT, Pereira MC, Soares CT, Fleury RN. Sporadic and multiple neurofibromas in the head and neck region: a retrospective study of 33 years. Clin Oral Investig. 2007;11:165–9. [PubMed]
21. Sahota JS, Viswanathan A, Nayak DR, Hazarika P. Giant neurofibroma of the tongue. Int J Pediatr Otorhinolaryngol. 1996;34:153–7. [PubMed].