Indian Journal of
Plastic Surgery

IJPS 2001; 34: 70-74

Partial Excision of Giant Plexiform Neurofibromatosis

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Summary

Seven cases of plexiform neurofibromatosis, which underwent single or multiple partial excisions from 1996-2000 were reviewed for quality of scar, aesthetic results and any other complication. Early results were satisfactory. Follow up period was inadequate to comment on the malignant degeneration at the surgery site.

Key words : Neurofibromatosis, Partial excision, Plexiform

Introduction

For convenience of description, plexiform neurofibromatosis that poses a challenging problem to a surgeon during complete excision will be referred as giant plexiform neurofibromatosis. In recent past emphasis is on complete excision[1], though many agree that it is technically difficult[2].

The authors feel that complete excision is not practical considering the morbidity, poor socioeconomic conditions of the patients, technical difficulties and lack of facilities in many centers. Due to cosmetic and functional reasons; in selected individuals, multiple limited excisions may be more beneficial in aesthetic, functional and psychological rehabilitation and hence; may improve the patient's quality of life.

Review of literature Phacomatoses (Neurocuteneous Syndromes) includes Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), Tuberous sclerosis, and Von hippel lindau syndrome. NF1 or Von Recling Hausens disease is characterized by benign tumors of peripheral nerves, composed of Schwann cells (endoneurium) and fibroblasts, and pigmented skin lesions called cafe au lait spots. It is caused by mutation of the tumor suppressor gene NF1 on chromosome 17q, which encodes for a protein *neurofibromin* (GTPase), which acts through the 'ras' pathway. NF2 is characterized by bilateral acoustic schwannomas, meningiomas, gliomas and schwannomas of cranial/spinal nerves[3].

Diagnostic criteria for NF1 includes: 1) six or more cafe '*au lait spots* >5mm in pre pubertal persons and >15mm in adults, 2) Two or more neurofibromas /one plexiform neurofibroma, 3) Freckling of axillary or inguinal regions, 4) Optic gliomas, 5) Osseous lesions (e.g. pseudo arthrosis /thinning of long bones, 6) Two or more Lisch nodules (hamartomas of iris) and 7) First degree relative with NF1. Presence of two or more of the above is diagnostic[1].

The plexiform variant is rare and occurs in connection with the branches of the fifth cranial nerve or the extremities[4]. However giant plexiform neurofibroma has been reported at various sites like chest[5], back[1], tongue[6], penis[7-8] and labia[9].

Morphologically these neoplasms involve subcu-

taneous tissue wherein nerves are tortuous and thickened, overlying skin is hyper pigmented and they may achieve massive proportions .The host nerve is irregularly expanded. Each fascicle is infiltrated by neoplasm so that the proximal and distal extents are not clear. Histologically they show a loose myxoid background with diminished cellularity .The cell phenotypes include Schwann cells with elongated nuclei and extension of pink cytoplasm; multipolar fibroblasts; a sprinkling of inflammatory cells. Hence they are similar to antoni - 2 type of schwannomas.[10]

The reported incidence[10] of NF1 is 1:3,000. The incidence[10] of malignancy in NF1 is 5-8%.

In children with NF1 surgical intervention is needed in approximately 20% of cases. In large plexiform neurofibroma often multiple resections are needed[11].

Giant plexiform neurofibromatosis of as much as 70X55cm, weighing up to 32 kg have been reported[1].

Current emphasis is on complete excision, which is technically difficult in a giant neurofibroma. Nahabedian[1] et al have advised meticulous preoperative planning with MRI, CT and arteriography to define the extent of the mass. In contrast complete excision may not be safe, may lead to considerable morbidity in many situations where the lesions are in proximity to important structures. Plexiform neurofibromas are notorious for their bleeding tendency and inability to achieve complete surgical excision due to extensive diffuse nature of the lesions without distinct natural tissue planes[2].

Methods

A retrospective analysis of 7cases of giant plexiform neurofibromatosis admitted in the Department of Plastic Surgery, Kasturba Hospital Manipal was done. Records of all these patients were reviewed and relevant data was collected (Table 1). All the patients were diagnosed clinically and proved on histopathological examination. These patients were followed from 6 months to 5 years.

Table 1. Tabular presentation of case summaries

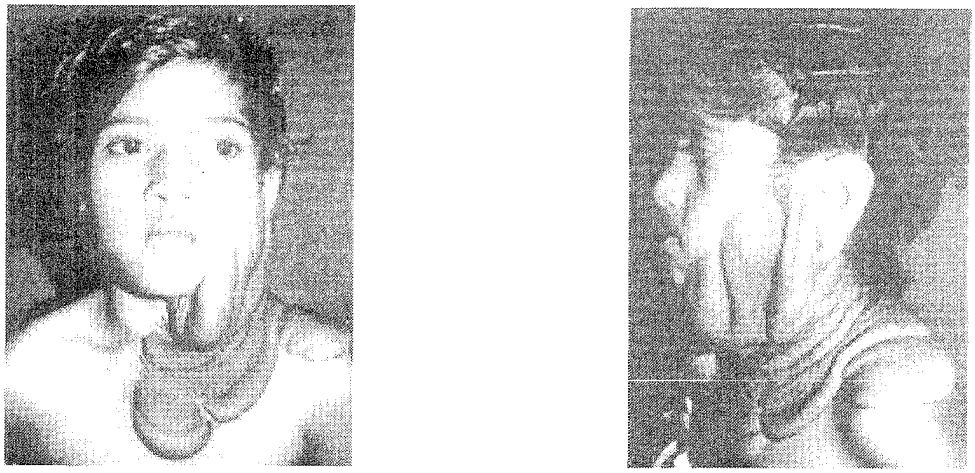
Sl. No.	Age (yrs)	Sex	Classification	site	Surgery	Hospitalization (Days)	Intraptranfusion Y/N	Postop Tranfusion Y/N	Postop Subjective evaluation
1	11	F	NF1	Chest Shoulder Neck Face	Partial excision 4 times	16, 16, 13 & 10	Y	Y	Satisfactory
2	17	M	NF1	Left lip ear & right eyelid	Partial excision	10	N	N	Satisfactory
3	22	M	NF1	Scalp, face and orbit	Partial excision	11	Y	Y	Satisfactory
4	20	F	NF1	Forehead	Partial excision	11	Y	Y	Satisfactory
5	18	F	NF1	Back	Partial excision	10	N	N	Satisfactory
6	30	M	NF1	Scalp	Partial excision	10	N	N	Satisfactory
7	25	M	NF1	Chest arm	Partial excision 2 times	14 & 11	Y	Y	Satisfactory

Operative details

Incision was made at the base of the lesion along the markings drawn after assessing the loose skin by pinching skin between index finger and thumb. Excessive bleeding was observed on cutting soft and friable skin and neurofibromatous tissue from relatively larger blood vessels present in the neurofibromatous tissue. Hemostasis, though difficult, was achieved by electro-coagulation or ligature (transfixation if required). Wound was closed in two layers after placing drain and finally pressure dressing was done.

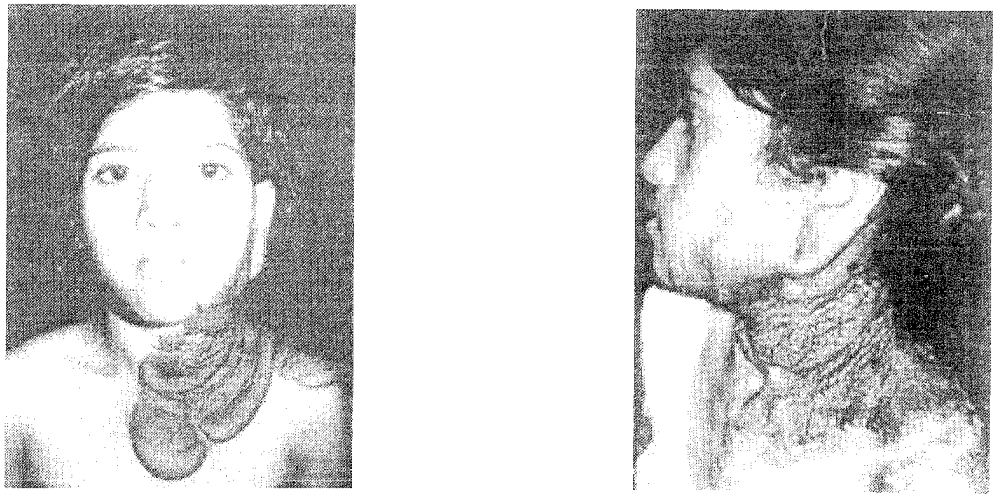
Results

As per patient's evaluation results were satisfactory both cosmetically and functionally in all the cases (Fig 1-6). Due to extensive nature of the lesion in two cases (Table 1 : case 1 and case 7) to reduce the blood loss and morbidity, partial excisions done two or more times. In both these cases first excision required blood transfusion intra-operative (one unit) and post- operatively (one unit), and hospital stay was more than two weeks. Subsequent limited excisions were done to avoid blood transfusion and reduce the total



(a) (b)

Fig 1. Pre-operative photograph of the patient (case 1) showing plexiform neurofibromatosis involving face, neck, chest wall and shoulder (a) Front view (b) side view



(a) (b)

Fig 2. Post-operative photograph of the patient shown in Fig 1. (a) front view after excision of facial lesions (b) side view after four excisions

Fig 3. Pre-operative photograph of patient (case 3) showing plexiform neurofibromatosis involving scalp, face and orbit

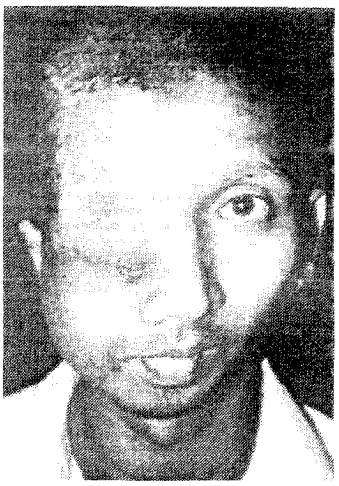


Fig 4. Post-operative photograph of the patient shown in Fig 3

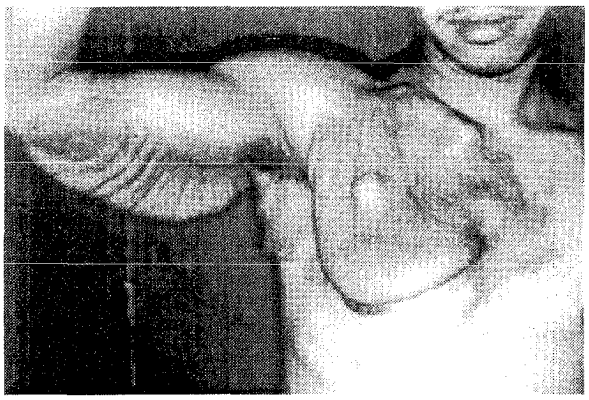


Fig 5.Pre-operative photograph of the patient (case 7) showing plexiform neurofibromatosis of chest wall and arm

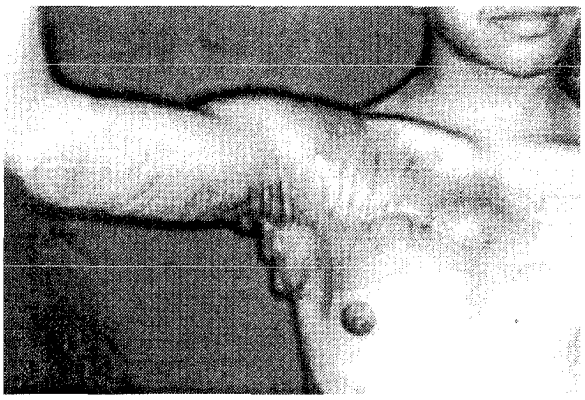


Fig 6. Post-operative photograph of the patient shown in Fig 5

hospital stay. In case 2, 4 and 6, partial excision was done due to cosmetic reasons. In case 2 and 6, total hospital stay was 10 days. In case3 and 4 blood transfusions were required and hospital stay was 11 days In all the cases drain was removed after 48 - 72 hours. All the scars were aesthetically acceptable and no recurrence or other complication was noticed during the follow up period.

Discussion

Extensive plexiform neurofibromatosis poses a challenging problem in our clinical practice. In all 7 cases under present study, complete excision was not possible either due to excessive intraoperative bleeding, extensive size of the lesion, close proximity to the important anatomical structures or possible unacceptable cosmetic result. On operation table, the neurofibromatosis tissue was found to be friable and blood vessels were relatively larger in size and bled profusely due to decreased ability to contract at the cut ends. Most of the cut ends of the vessels failed to stop bleeding even after electrocautery and required ligation or running sutures. After analyzing the results in terms of improved cosmetic results and less morbidity, the authors recommend multiple partial excisions in suitable cases as preferred method of excision. However, patients should be

followed for longer duration for any recurrence and malignant degeneration, and informed for self-examination and should be report immediately if required.

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