Loco-regional Fasciocutaneous Flaps for Prompt Repair of Type I Neurofibromatosis*

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ABSTRACT

This paper aims to study surgical techniques, whether associated or isolated, which provide less stigmatizing and deforming results for patients with type I neurofibromatosis (NF I), in an attempt to ensure a quality of life with more dignity and better social integration. Thirty patients with NF I were studied for a period of six years. Different surgical techniques were used to remove lesions ranging from 3 to 51 cm, located in different anatomical regions, and their results were analyzed. Twenty-one patients benefited from excisions and repairs with flaps, and the remaining, from procedures of excision and suture or z-plasty. Loco-regional fasciocutaneous flaps provided the best results both functionally and esthetically, with fewer sequelae. Even in the cases with residual sequelae in the skin used for the cutaneous flaps, the disease did not evolve and there was no malignization. Loco-regional fasciocutaneous flaps enabled the repair of wide bleeding areas and the resection of larger tumors. Medium-sized lesions are satisfactorily removed using the s-plasty ou z-plasty techniques for coverage. Small lesions are appropriately repaired using excision and suture, with no need for flaps, especially those located on the face and nose.

INTRODUCTION

Neurofibromatosis or phakomatosis, a word of Greek origin meaning a spot, was introduced in the medical literature in 1768. The study of neurofibromatosis goes back to the seventeenth century when the surgeon Mark Akenside described, in a 60 year-old man, multiple cutaneous and subcutaneous tumors that he attributed to paternal heritage[1]. Toselius (1793) and Smith (1849) reported findings in two similar cases by using anatomical and histopathological necropsy studies[1]. However, half of NF I cases are sporadic, and parents are not affected, with the disease appearing spontaneously, as a consequence of genetic alterations or mutations[1].

It was Friedrick von Recklinghausen, in 1882, also based on necropsy findings, who best described this pathology and clarified its origin in the Nervous System with cutaneous, visceral and neurological consequences. For that reason, it was called neurofibromatosis or Von Recklinghausen's disease[1].

NF I is a family genetic abnormality characterized by the growth of tissues derived from neural crests; its changes manifest themselves before the thirtieth day of intra-uterine life, when visceral differentiation starts[2].

The gene that causes NF I is located next to the centromer of the long arm of chromosome 17. The disease represents a new mutation in fifty percent of the patients affected.[3]. The proteins that cause these changes were identified and named neurofibromin for NF I, and Merlin protein or schwannomin for NF II, respectively[4].

TABLE I

Gender	Age	Skin Color	Site of Lesion	Characteristics of Lesions	Surgical Technique
F	16	Brown	Leg	Lobulated, gelatinous, brown	Fasciocutaneous flap
F	16	Brown	Leg	Wrinkled, gelatinous, brown	Fasciocutaneous flap
M	29	Brown	Face	Wrinkled, gelatinous, brown	Fasciocutaneous flap
M	29	Brown	Face	Wrinkled, gelatinous, brown	Fasciocutaneous flap
M	29	Brown	Face	Wrinkled, brown	Fasciocutaneous flap
F	15	Black	Leg	Lobulated, gelatinous, brown	Fasciocutaneous flap
F	15	Black	Leg	Lobulated, brown	Fasciocutaneous flap
F	15	Black	Leg	Wrinkled	Fasciocutaneous flap
F	48	Brown	Face	Bulging, pilose, gelatinous	Fasciocutaneous flap
F	48	Brown	Face	Wrinkled, pilose	Fasciocutaneous flap
M	51	White	Thigh left	Bulging, hardened	Fasciocutaneous flap
M	51	White	Buttocks, legs	Gelatinous, bulging	S-plasty
M	17	Brown	Face	Gelatinous, bulging, brown	Fasciocutaneous flap
M	17	Brown	Face	Bulging, brown	Fasciocutaneous flap
M	27	White	Head	Wrinkled, hardened	Fasciocutaneous flap
M	27	White	Thorax	Wrinkled, gelatinous	Fasciocutaneous flap,
F	28	White	Genitals	Lobulated, hardened	Excision/bloody zone
F	28	White	Genitals	Granulated tissue, redish	Fasciocutaneous flap.
M	31	Brown	Face, buttocks, arm	Bulging, gelatinous	Z-plasty
M	8	Brown	Eyebrow	Bulging, gelatinous	Excision suture
M	45	White	Arm, dorsum	Bulging, gelatinous	Z-plasty
M	52	White	Upper limb	Bulging, gelatinous	Z-plasty
F	58	White	Face	Hardened, wrinkled	Z-plasty
F	58	White	Face	Hardened, wrinkled	Fasciocutaneous flap
F	58	White	Face	Hardened, wrinkled	Fasciocutaneous flap
F	58	White	Face	Hardened, wrinkled	Skin and mucosa graft
F	18	White	Hand	Hardened, bulging	Fasciocutaneous flap
M	9	Brown	Eyebrow	Bulging, gelatinous	Excision and suture
F	11	Brown	Eyebrow	Bulging, gelatinous	Excision and suture
F	39	Black	Face	Bulging, gelatinous	Z-plasty
M	60	Brown	Thorax	Bulging, hardened	Fasciocutaneous flap
M	60	Brown	Upper limb	Gelatinous, bulging	Graft skin
F	27	Brown	Dorsum	Gelatinous, bulging	Z-plasty
F	27	Brown	Dorsum	Gelatinous, bulging	Excision and suture
M	14	Brown	Buttocks	Bulging, hardened	Fasciocutaneous flap
M	14	Brown	Right thigh	Bulging, hardened	Z-plasty
F	27	Brown	Face	Gelatinous, bulging	Fasciocutaneous flap
F	27	Brown	Upper limb	Bulging, hardened	Z-plasty
F	23	White	Thorax	Bulging, hardened	Fasciocutaneous flap
M	20	Brown	Face	Bulging, gelatinous	Fasciocutaneous flap
M	20	Brown	Face	Hardened, bulging	Z-plasty
M	20	Brown	Dorsum	Gelatinous, bulging	Fasciocutaneous flap
M	20	Brown	Upper limb	Hardened, bulging	Z-plasty
M	48	Brown	Thorax	Gelatinous, bulging	Z-plasty
M	47	White	Face	Wrinkled, hardened	Fasciocutaneous flap
F	56	Black	Face	Lobulated, hardened	Fasciocutaneous flap
M	45	White	Face	Wrinkled, gelatinous	Fasciocutaneous flap
M	45	White	Upper limb	Hardened, bulging	Z-plasty
F	50	White	Head	Hardened, bulging	Z-plasty
M	42	Brown	Thorax	Hardened, bulging	Z-plasty
M	42	Brown	Upper limb	Hardened, bulging	S-plasty
M	20	Black	Face	Gelatinous, bulging	Fasciocutaneous flap

Lesion sites and characteristics, and surgical techniques.

Other factors account for tumor growth, including androgen and estrogen factors in the passage into puberty, and the increase in hormone loads during pregnancy, facts which have not been proven so far[5].

The growth of tumors in NF I is unpredictable, with periods of rapid growth and others of slower growth; many remain static indefinitely after a quick development. This irregular behavior makes it difficult to document the effectiveness of a potentially healing treatment[3].

Therapy is limited to surgical excisions that are often unsatisfactory, because in many cases the full resection of large lesions is rarely possible and tumors emerge continuously over the lifetime[6]. The existing literature is generally limited to isolated reports, which are disseminated to the medical community only because they attract attention due to the deformities[1,5,7-15].

From the surgical standpoint, there are few current publications seeking to improve the deformation and disfigurement of patients who suffer from this genetic abnormality.

The goal of this study was to present associated or isolated surgical techniques, in an attempt, through less traumatic and more effective surgeries, to achieve less stigmatizing and deforming results and a quality of life with more dignity and more rapid social reintegration.

METHODS

Thirty patients (17 men, 13 women), with ages rang-

Table II

Gender	Age	Skin Color	Site of lesion	Characteristics of lesions	Surgical technique	Size (cm)	Weight (g)
F	16	Brown	leg	lobulated, gelatinous, brown	fasciocutaneous flap	32,0 x 12,0	1.120
F	16	Brown	leg	wrinkled, gelatinous, brown	fasciocutaneous flap	28,0 x 6,0	230
M	29	Brown	face	wrinkled, gelatinous, brown	fasciocutaneous flap	13,5 x 9,0	180
M	29	Brown	face	wrinkled, gelatinous, brown	fasciocutaneous flap	15,0 x 6,5	200
M	29	Brown	face	wrinkled, brown	fasciocutaneous flap	10,0 x 3,5	55
F	15	Black	leg	lobulated, gelatinous, brown	fasciocutaneous flap	36,0 x 10,0	745
F	15	Black	leg	lobulated, brown	fasciocutaneous flap	30,0 x 6,0	240
F	15	Black	leg	wrinkled	fasciocutaneous flap	25,0 x 4,0	110
F	48	Brown	face	bulging, pilose, gelatinous	fasciocutaneous flap	12,0 x 7,0	210
F	48	Brown	face	wrinkled, pilose	fasciocutaneous flap	8,0 x 4,0	80
M	51	White	left thigh	bulging, hardened	fasciocutaneous flap	15,0 x 12,0	380
M	17	Brown	face	gelatinous, bulging, brown	fasciocutaneous flap	11,0 x 5,5	120
M	17	Brown	face	bulging, brown	fasciocutaneous flap	6,0 x 3,5	65
M	27	White	head	wrinkled, hardened	fasciocutaneous flap	51,0 x 28,0	1.800
M	27	White	thorax	wrinkled, gelatinous	fasciocutaneous flap	30,0 x 15,0	900
F	28	White	genitals	Granulated tissue, redish	fasciocutaneous flap	23,0 x 19,0	850
F	58	White	face	hardened, wrinkled	fasciocutaneous flap	6,0 x 2,0	30
F	58	White	face	hardened, wrinkled	fasciocutaneous flap	4,5 x 3,5	45
F	18	White	hand	hardened, bulging	fasciocutaneous flap	3,0 x 2,0	15
M	60	Brown	thorax	bulging, hardened	fasciocutaneous flap	12,0 x 9,0	110
M	14	Brown	buttocks	bulging, hardened	fasciocutaneous flap	8,0 x 7,0	180
F	27	Brown	face	gelatinous, bulging	fasciocutaneous flap	10,0 x 6,0	70
F	23	White	thorax	bulging, hardened	fasciocutaneous flap	16,0 x 12,0	245
M	20	Brown	face	bulging, gelatinous	fasciocutaneous flap	15,0 x 10,0	230
M	20	Brown	dorsum	gelatinous, bulging	fasciocutaneous flap	12,0 x 6,0	140
M	47	White	face	wrinkled, hardened	fasciocutaneous flap	10,0 x 8,0	60
F	56	Black	face	lobulated, hardened	fasciocutaneous flap	15,0 x 8,0	110
M	45	White	face	wrinkled, gelatinous	fasciocutaneous flap	8,0 x 5,0	80
M	20	Black	face	gelatinous, bulging	fasciocutaneous flap	12,0 x 6,5	100

Fasciocutaneous flaps.

ing from 08 to 60 years of age were selected for this study.

All patients were previously assessed, so as to ensure that all of them could undergo the proposed surgeries. Surgical incisions were previously marked using methylene blue, with which the surgeon also drew flaps, z-plasties, and s-plasties, the zig-zag incisions that were meant to guide surgical closing, as well as to assess the extension of resections and the technique that was being used.

Considering the rich vascularization, in the cases of head and neck tumors, previous infiltration was performed with 0.5% lidocaine hydrochloride in association with 1:400,000 epinephrine, as well as preventive cerclage with separated stitches, with the aim of reducing bleeding. For upper and lower limb tumors, it was imperative to use a pneumatic tourniquet.

In all patients with encapsulated and well-individualized tumors that could be easily enucleated, the cutaneous lining was dissected in its entire thickness with a digital dissection. For tumors affecting the skin, 0.5 cm of its thickness were preserved for making the flap that served to repair the bloody zone left by the resection. When lesions were located in a plane between the fascia and muscles, they did not offer difficulty for exeresis, which ensured stitches would not leave motor or sensory sequelae. After tumor removal, hemostasia was reviewed, and the muscular fascia was sutured, if it had been lacerated or incised, with a 3-0 nylon monofilament suture. Next, excesses of the fasciocutaneous flap were excised, vessels were clotted, the pneumatic tourniquet was removed, in the case of lower and upper limbs, or the cerclage, when the procedure had been on the cephalic segment. The following phases consisted of closing the subcutaneous cellular tissue with 3-0 absorbable monofilament suture and the skin was sutured with separate stitches, interspersed with continuous 4-0 nylon monofilament sutures. As operated areas left a bleeding zone and a virtual dead space, a continuous suction drain was placed and removed when drainage stopped or reached 20 and 50 ml in 24 hours.

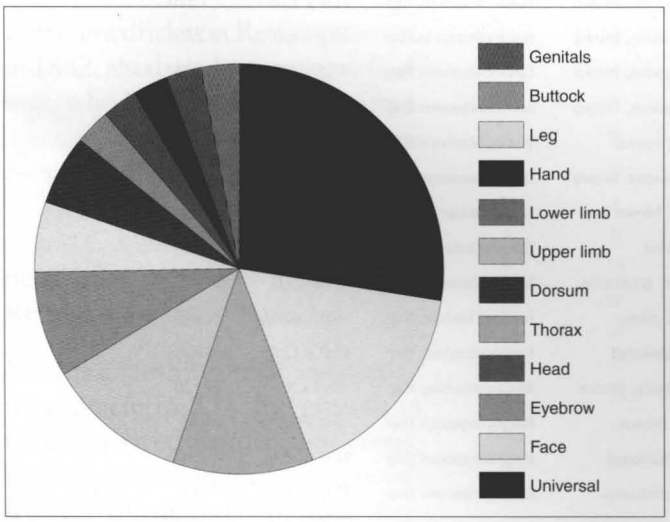

Fig. 1 – Anatomical distribution of lesions.

Dressings were made with padded gauze, simple gauze, crepe bandages, tailor-made elastic meshes or masks with moderate compression, which were removed every two days.

All patients were observed and followed up for 6 years (1995 to 2001). Revisions were made on a monthly basis, in the first 6 months and, from then on, biannually. The quality of scars, evolution and emergence of new tumors, pain and signs of relapse were assessed.

Table I lists the information about all patients, site and characteristics of lesions and the surgical techniques used in the 52 resections.

RESULTS

In the period between March 1996 to March 2001, thirty patients diagnosed with NF I underwent tumor resections, 21 of which benefited from the use of loco-regional fasciocutaneous flaps. Table II shows the information of patients, characteristics of lesions, surgical techniques, size and weight of surgical specimens.

There was universal impairment in

Table III

Characteristics of lesions	GenderM	Gender F	Total	%
Café-au-lait spot	20	10	30	44,1
Freckles	12	6	18	26,5
Plexiform tumor	1	2	3	4,4
Hamartoma (Lisch nodule)	2	0	2	2,9
Changes in the zygomatic arch	4	2	6	8,8
Cortical thickening	0	2	2	2,9
Scoliosis	4	3	7	10,3
Totals	43	25	68	100,0

Classical signs of NF1.

10 cases; upper limbs accounted for 05: 04 of the forearm and arm and 01 on the hand; the cephalic segment accounted for 10: 06 on the face, 01 on the head and 03 on the eyebrow; there were 03 in lower limbs, of which 01 was on the thigh and 02 on the leg; 01 on the buttocks; 04 on the thorax, dorsum 02 and genitals 01. Percentages and their sites are shown in Figure 1.

The classical signs of NF I, as well as other lesions that may comprise the picture, are presented in Table III, where it is possible to see the incidence and the percentages found in the research.

There were plexiform neurofibromas in four cases and in all of them results were considered very good, esthetically, physiologically, and functionally (Figs. 2 to 6).

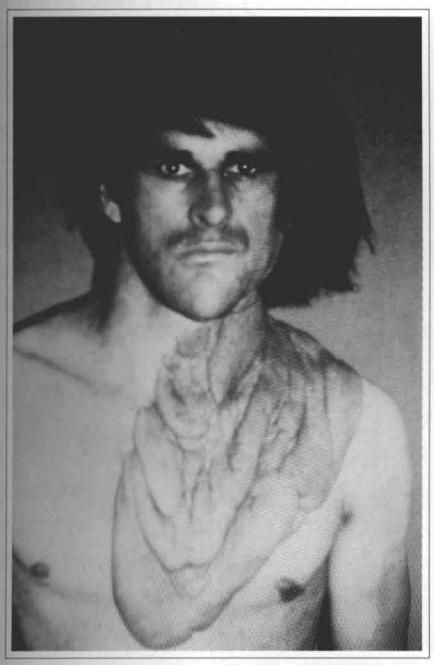

Fig. 2 – Plexiform neurofibroma on the thorax.

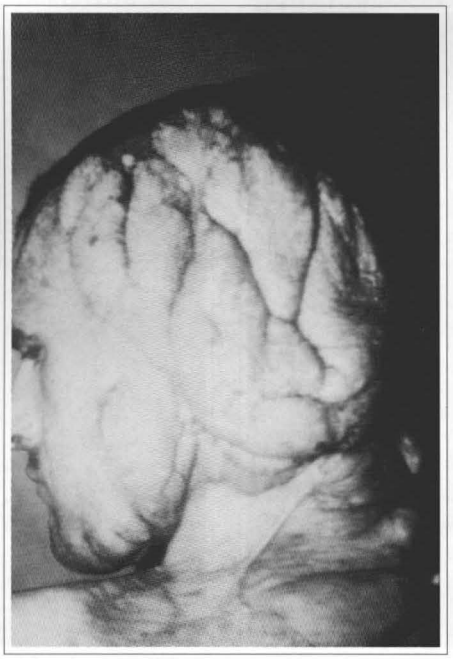

Fig. 3 – Plexiform neurofibroma in the craniocervical area.

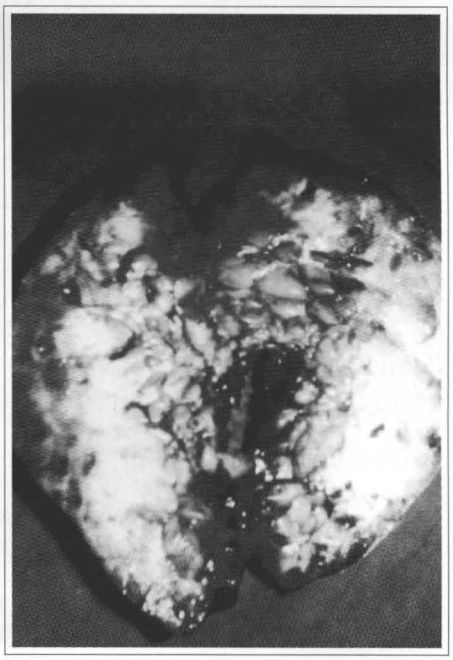

Fig. 4 – Extirpated neurofibromas in the craniocervical region.

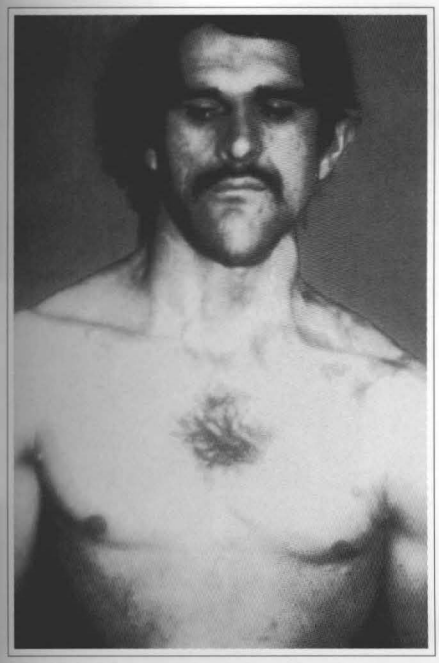

Fig. 5 –Frontal view of final result after 2 years.

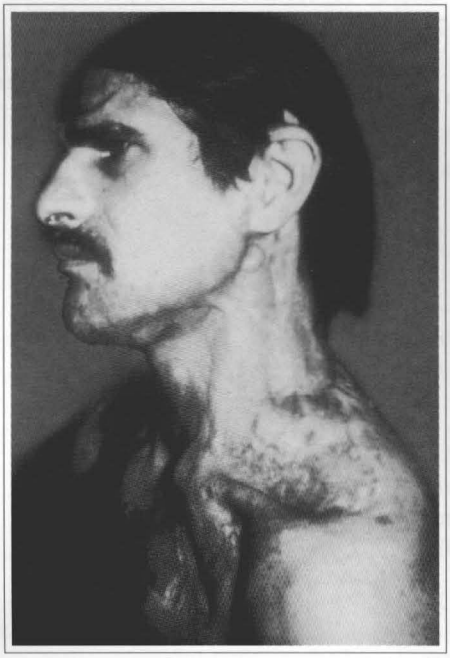

Fig. 6 – Left lateral view of final result after 2 years.

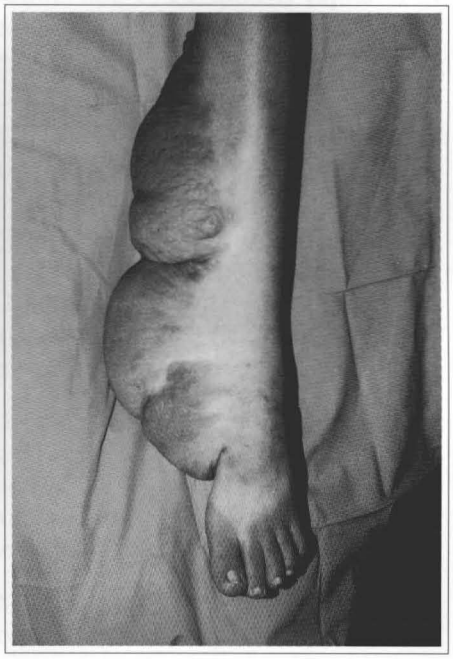

Fig. 7 – Leg left with plexiform neurofibroma; view of tumor.

There were three cases in the lower limbs, 1 on the thigh and 2 on the leg, one of them can be assessed in figures 7 to 9. One rare case, affecting the genital area, is shown in figures 10 to 13, in which the tumor affected the suprapubic, pubian and perineal regions.

In the 30 patients the evaluation of scars showed: 18 very good, 9 good, and 3 satisfactory. Pain disappeared in the operated regions in all cases. There was no dehiscence, tissue suffering, infection or necrosis in any of the cases. Scars formed, on average, in 21 days. New tumors were not noticed and there were no relapses.

Of the school age patients, 9 went back to educational activities; among adults, 12 men and 6 women went back to work, and 28 reintegrated into social life.

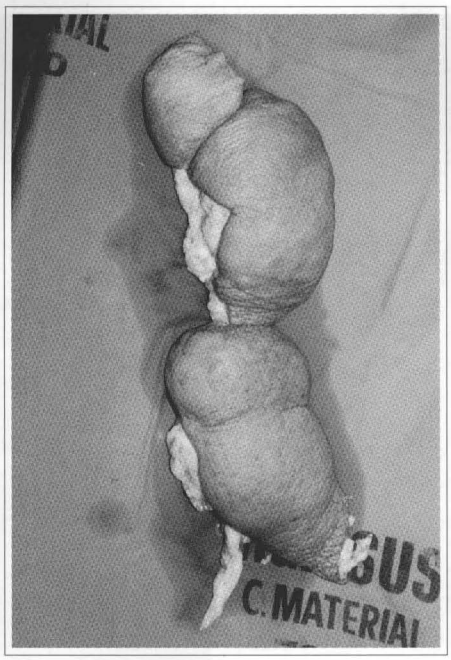

Fig. 8 – Leg left with plexiform neurofibroma; tumor excised.

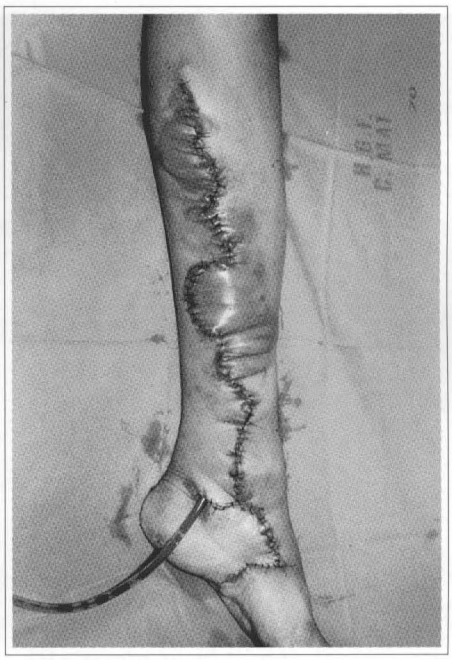

Fig. 9 – Leg left with plexiform neurofibroma; transoperative result.

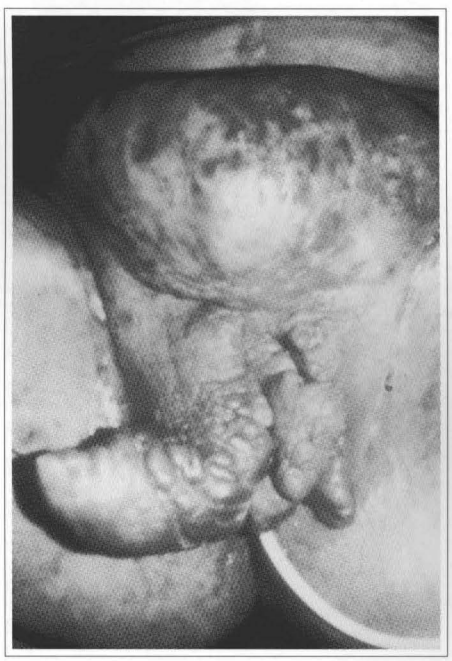

Fig. 10 – Pubian and genitocrural plexiform neurofibroma.

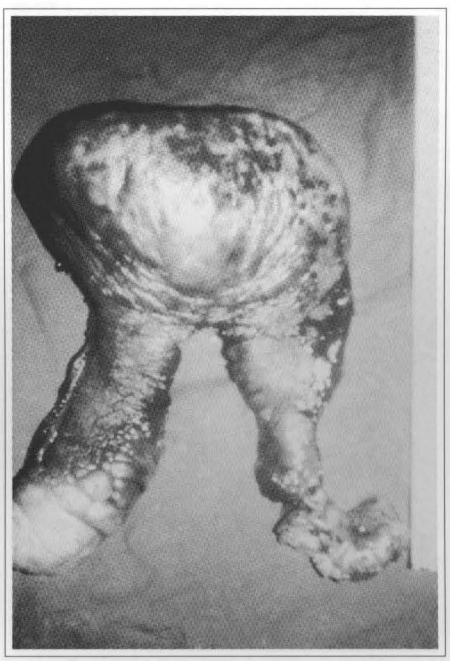

Fig. 11 – Pubian plexiform neurofibroma; extirpated tumor.

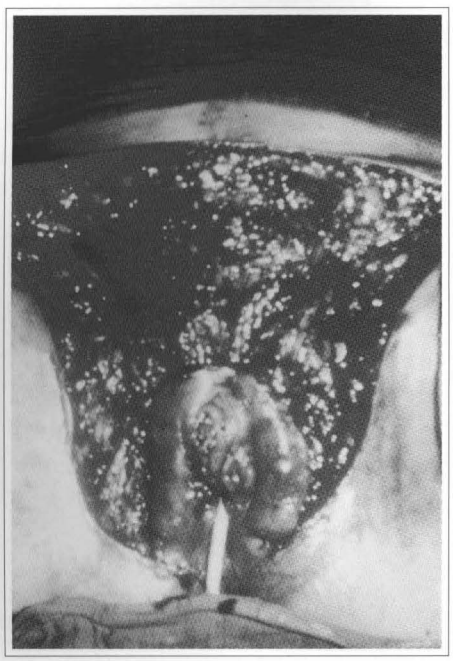

Fig. 12 – Pubian plexiform neurofibroma; bloody zone with transoperative result.

DISCUSSION

The existence of multiple neurofibromas is one of the defined hallmarks of NF I. Von Recklinghausen's classical description, in 1882, associating NF I to peripheral nerves prevailed for two centuries, and clinical observations provided many indications for the understanding of this pathology.

Papers published by H. Roger (1901), on nervous tumors, and Hosoi (1901), about the malignant degeneration enhanced research.

The frequency of malignization is difficult to be calculated. Daniel, Steward and Capelland (1882), cited by Baudet *et al.* (1976), believed that surgical intervention would serve as an accelerating factor for malignancy[6]; d'Agostinho, Soule and Miller (1963), cited by the same author, claim that sarcomas are often adjacent to neurofibromas[6].

This research did not detect malignization in any of the 52 specimens sent to the laboratory for histopathological examination.

Pitanguy *et al.* (1983) quote 23 cases about which they say: "Treatment consists of resecting lesions that affect function and/or aesthetics, using surgical techniques so as to correct the deformity", without, however, describing the details of the techniques used, or any postoperative result, merely showing preoperative photos[9].

The present study demonstrated that it is possible to repair large bleeding areas (largest 51.0 x 28.0 cm), with an average of 16.4 x 8.1 cm, resulting from the exeresis of large tumors (largest 1,800 g and average 300 g), using loco-regional fasciocutaneous flaps, thus providing adequate cutaneous coverage, given that simple excision, without cutaneous flaps, would make their surgical excision impossible.

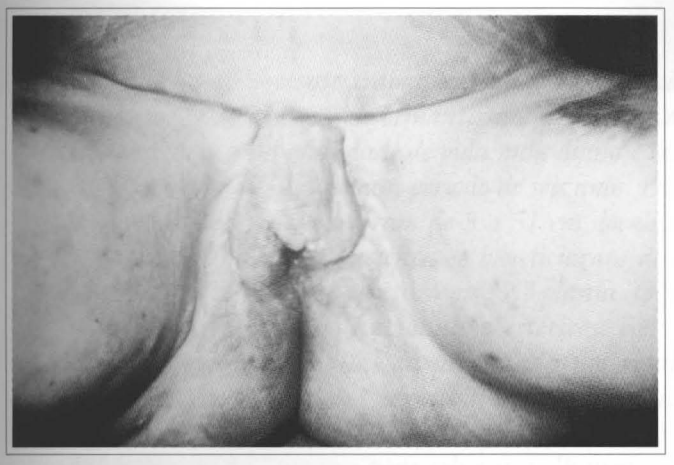

Fig. 13 – Pubian plexiform neurofibroma; result after 18 months.

The treatment suggested by many authors is limited to resecting tumors, especially those that affect mental and aesthetic functions, or when they cause pain, discomfort, functional incapacity, or present accelerated growth leading to suspicion of malignant transformation[5,8,9,12,13,15].

Considering that most NF I tumors are benign lesions, it is not justifiable to surgically extirpate noble tissues, like motor and sensory nerves, eyelids and conjunctiva, orbital exenteration or facial bones[14]. Multiple subtotal resections are recommended, so as to avoid futile, hasty interventions aimed at removing lesions totally[15].

Excision and suture and small skin grafts used on smaller lesions and on special sites, like face and nose, permitted complete cutaneous coverage.

S-plasty (average 8.5 x 5.5 cm and 135 g) and z-plasty (for lesions up to 8.5 x 4.8 cm and 82.6 g) proved to be effective in the repair of medium-sized lesions, for which they are one of the choices.

CONCLUSIONS

Loco-regional fasciocutaneous flaps make it possible to resect larger tumors and to adequately repair large bloody areas. Medium-sized lesions can be satisfactorily removed with perfect cutaneous coverage, using either s-plasty or o z-plasty. Small lesions can be promptly resected with no need of flaps, except for lesions on the face and nose.

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