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CASE REPORT

Electrosurgical excision technique for the treatment of multiple cutaneous lesions in neurofibromatosis type I

Steven M. Levine^a, Elie Levine^b, Peter J. Taub^b, Hubert Weinberg^{b,*}

^a New York University School of Medicine, 550 First Avenue, New York, NY 10016, USA

^b Division of Plastic Surgery at Mount Sinai School of Medicine, 1 Gustave L. Levy Place, New York, NY 10029, USA

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Summary Neurofibromatosis I (NF1) is an autosomal dominant disease that presents with multiple cutaneous lesions often numbering into the 500–1000 range. In addition to the psychosocial implications, there are limited surgical options for this condition. A series of 97 consecutive patients with NF1 presented with numerous cutaneous lesions. Treatment involved electrocautery excision of the lesions using a handheld device in one or more stages. This technique resulted in the removal of large numbers of lesions in limited stages, with minimal scarring, minor discomfort and high patient acceptance. For patients with NF1, single or multistage excision of cutaneous lesions using electrocautery produces an acceptable aesthetic result with high patient satisfaction.

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Neurofibromatosis I (NF1), or van Recklinghausen disease, is seen in approximately 1 in 2600 to 1 in 3000 live births.¹ It is an autosomal dominant genetic disorder with complete penetrance, yet despite this the ultimate expression is highly variable.^{2,3} The criteria for diagnosing NF1 include: (1) six or more café-au-lait spots >5 mm in prepubertal persons (>15 mm in postpubertal persons), (2) two more neurofibromas of any type, or one plexiform neurofibroma,

(3) freckling of the axillary or inguinal regions, (4) optic glioma, (5) two or more Lisch nodules, (6) osseous lesions such as sphenoid dysplasia and thinning of long bone (with or without pseudoarthrosis), and (7) a first-degree relative (parent, sibling, offspring) with NF1. A definitive diagnosis requires two or more criteria to be present.⁴

Patients with NF1 are prone to develop numerous sequelae including optic glioma, intracranial tumours, kyphoscoliosis, tibial bowing, language and learning delay, hypertension, leukaemia, and sarcoma. They also report cosmetic disfigurement as a result of the growth of numerous benign connective tissue tumours. These lesions are comprised of a mixture of Schwann cells, fibroblasts

* Corresponding author. 1050 Park Avenue, New York, NY 10028, USA. Tel.: +1 917 492 4200; fax: +1 917 492 4300.

E-mail address: hubert.weinberg@mssm.edu (H. Weinberg).

and mast cells.⁵ Four types of neurofibromatous lesions exist: cutaneous, subcutaneous, nodular plexiform and diffuse plexiform. Cutaneous neurofibromas are the most common type and consist of soft connective tissue arising from cells in the peripheral nerve sheath.⁶

The lesions in NF1 typically appear prior to or during adolescence and increase in size and number with age. They can vary in number from just a few to several thousand, with the highest density occurring over the trunk. Local pruritus may be associated with accelerated tumour growth and emotional distress.

Subcutaneous neurofibromas are similar to their cutaneous counterparts and usually become apparent at the start of adolescence or early adulthood. These lesions present as firm, tender nodules along the course of peripheral nerves. Both the cutaneous and subcutaneous types are benign and do not carry an increased risk of malignant transformation.⁷ The nodular and diffuse plexiform neurofibromas become symptomatic because of their enormous growth potential and potential to stimulate underlying bone or compress surrounding tissues. In contrast to the benign cutaneous and subcutaneous forms, plexiform neurofibromas harbor a 5% lifetime risk of transformation into malignant peripheral nerve-sheath tumours.

Patients and methods

A retrospective review of 97 patients with NF1 and numerous, cutaneous lesions was undertaken. Each of the patients underwent at least one session of lesion excision by electrocautery by the senior author (HW).

The surgical technique was nearly identical in all cases. All procedures were performed on an ambulatory basis. A preoperative dose of intravenous cefazolin was given in all cases of nonpenicillin-allergic patients. The patient's skin was prepared with benzalkonium for antisepsis. One or two surgeons operated simultaneously with separate hand pieces utilizing E-Z Clean[®] needle-point tips (Megadyne, Draper, UT, USA), blend setting and the lowest effective current. Electrocautery was applied directly to the sessile lesions, essentially vaporizing them. In this case, no specimen was available for submission. Pedunculated lesions were grasped with a sterile forceps and excised at the base using the electrocautery tip. Here, a specimen was available for histopathologic examination. Subcutaneous components required deeper destruction, either by insertion of the needle point directly into the lesion or by applying digital pressure on the surrounding tissue to extrude the lesion. Rarely, a suture was required to close defects larger than 1 cm. The wounds were dressed with topical bacitracin ointment and patients given a prescription for a 4-day course of cephalixin if not allergic to penicillin. The bacitracin ointment was used twice daily for a period of five days. Showers were permitted on postoperative day 2.

Case reports

Case 1

A 47-year-old female with NF1 presented with numerous cutaneous lesions of face, trunk, arms and legs (Figure 1a

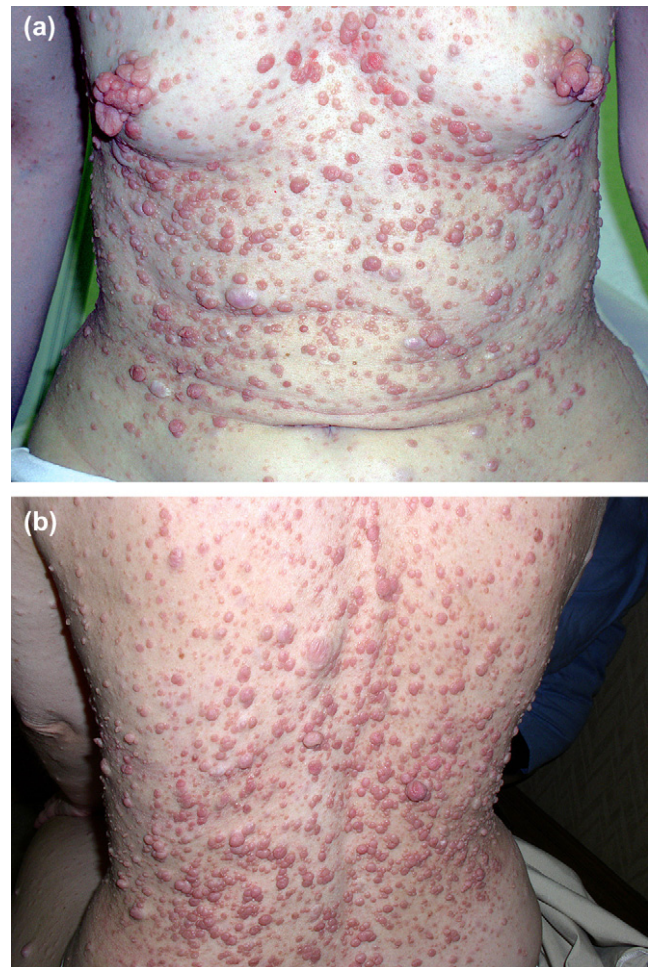


Figure 1 Preoperative photographs of case 1 demonstrating numerous cutaneous lesions of the (a) chest, specifically the nipple–areola complex, and (b) posterior trunk related to NF1.

and b). She initially underwent electrosurgical excision of the lesions on her posterior trunk. Six months later, she underwent a similar excision for the remaining lesions on her chest and abdomen. On follow up, she was noted to have no recurrence of the lesions and was content with the cosmetic result (Figure 2a and b).

Case 2

A 49-year-old female with NFI presented with extensive cutaneous involvement of the face, trunk and extremities (Figure 3). She was only interested in excising those lesions on exposed surfaces. Electrosurgical excision of the lesions covering her face and chest was performed in a single stage. On follow up, she was noted to have no recurrence of the lesions and was content with the cosmetic result (Figure 4).

Case 3

A 55-year-old female with NFI presented with cutaneous involvement of the face, anterior and posterior trunk, as well as extremities (Figure 5a and b). She underwent six separate electrosurgical excision procedures. All of the

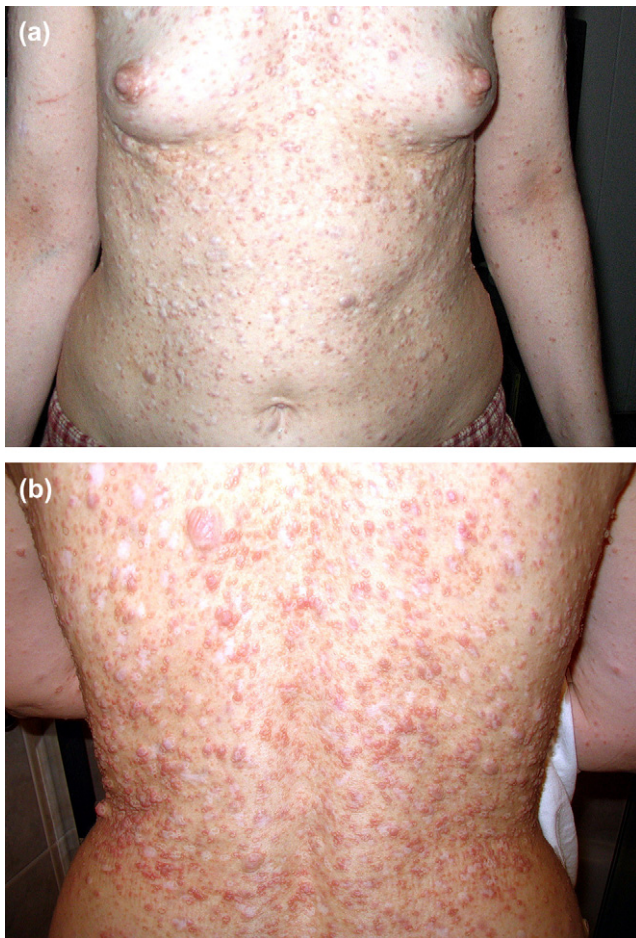


Figure 2 Six-month postoperative photographs of case 1 demonstrating improved appearance of (a) chest and breast, and (b) posterior trunk following electrocautery of cutaneous lesions.

sites were noted to have healed well on long-term post-operative follow up. On follow up, she was noted to have no recurrence of the lesions and was content with the cosmetic result (Figure 6a and b).

Results

Patients in the study ranged from 17 to 68 years of age. A total of 175 procedures were performed on the 97 patients. Fifty-nine patients were operated on in a single stage, whereas 23 patients underwent excision in two stages, and 15 underwent more than two procedures for an average of 1.8 procedures per patient to excise the majority of their lesions. The average number of lesions excised per session was 450. Surgical treatments were typically confined to within one or two zones of the body: the anterior trunk, arms, legs and face; or the posterior trunk, nape of neck and posterior extremities. The average length of surgery was 2 h. Postoperatively, patients were initially seen on a weekly basis, and healing time on the face was noted to be less than 10 days while that on the trunk and extremities was noted to be approximately 2–3 weeks.

All 97 patients were satisfied with the results as determined by direct questioning at the time of each

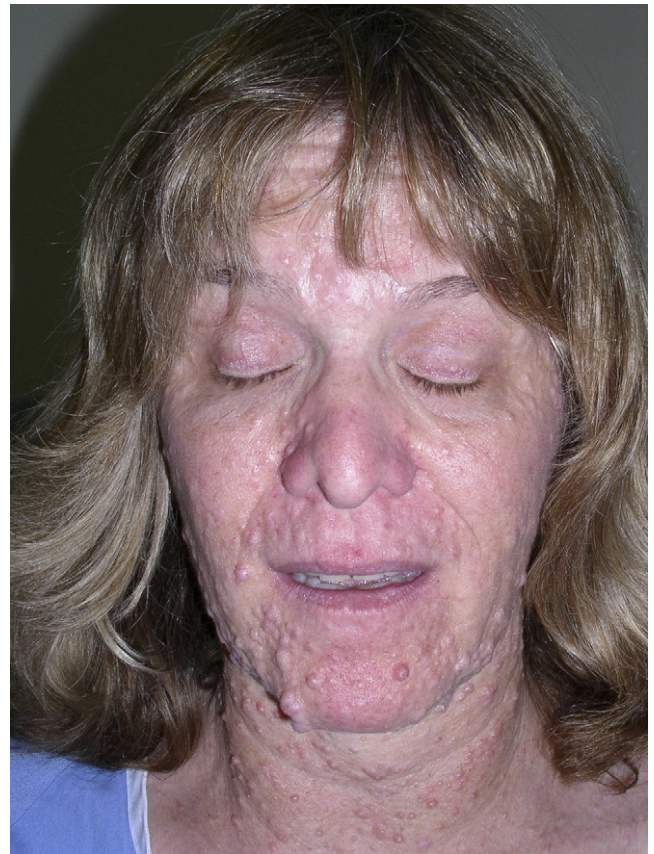


Figure 3 Preoperative photograph of case 2 demonstrating numerous cutaneous lesions of the face related to NF1.



Figure 4 Six-month postoperative photograph of case 2 demonstrating improved cosmesis of the face following electrocautery of cutaneous lesions.



Figure 5 Preoperative photographs of case 3 demonstrating numerous cutaneous lesions of the (a) face and (b) chest and breast related to NF1.

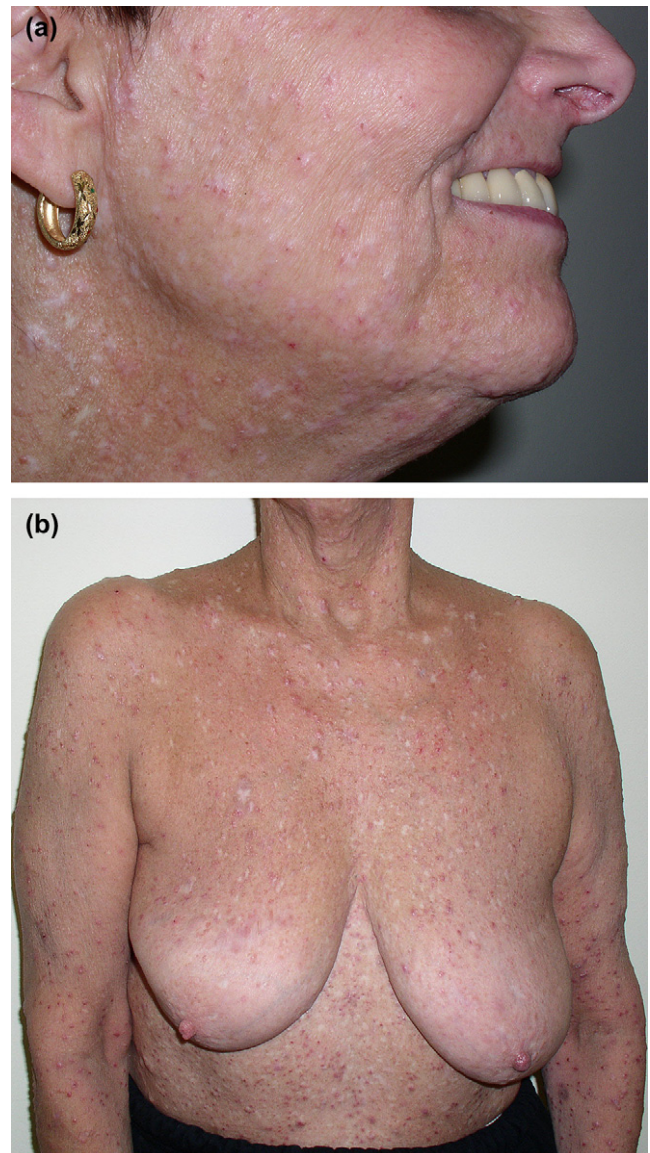


Figure 6 Six-month postoperative photographs of case 3 demonstrating improved cosmesis of the (a) face and (b) chest and breast following electrocautery of cutaneous lesions.

postoperative visit. The patients reported minimal discomfort during the immediate postoperative period and virtually no pain thereafter. Long-term results demonstrated minimal scarring with this technique.

Discussion

Since one of the most devastating consequences of NF1 is the psychosocial impact of the cutaneous lesions,^{8,9} removal can be of immeasurable benefit to the patient. Approximately half of NF1 patients have cutaneous tumours that number well into the hundreds.¹⁰ Lesions that occur around the face, neck and scalp account for roughly 48% of the total and are highly visible.¹¹ Those on the remainder of the body are most common on the trunk (64%), followed by the upper (54%) and lower (31%) extremities. In

addition to the skeletal deformities, seizure disorders and increased risk of certain cancers, patients are aware that there is a high likelihood that they will pass their disease on to their children. As a result, they can become socially withdrawn. Their cutaneous neurofibromas serve as a constant visible reminder of the perceived problems. Fortunately, these lesions can be removed with excellent cosmetic results.

There are several established techniques for removal of the lesions in NF1: surgical excision¹⁰ and/or CO₂ laser ablation.^{11,12} Surgical excision is a time-tested method that yields a fairly predictable scar, but removal of hundreds of lesions is impractical due to time constraints. This reality forces patients to choose a small subset of their lesions to be removed at one time, producing suboptimal patient satisfaction.

The described method for the treatment of multiple cutaneous neurofibromas has numerous benefits. Needle-point tip cautery provides instant haemostasis with minimal thermal damage to surrounding tissue. The technique is able to treat >500 lesions at any one sitting, in part due to the possibility of using two or more cautery devices simultaneously. The only similar published method to date used a single 1.5-cm monopolar diathermy wire loop.¹³ Under local anaesthesia, the neurofibroma was lifted with forceps and passed through the diathermy loop. Haemostasis was readily obtained and the wound was allowed to heal by secondary intention. Cosmesis was described as good, with re-epithelialization normally complete by three weeks. The authors described treating up to 250 lesions in 1 h using this technique.

Alternatively, CO₂ laser vaporization allows for treatment of numerous, widespread lesions under local anaesthesia. It also provides immediate haemostasis and permits healing via secondary intention with excellent cosmesis (usually a flat, smooth, depigmented scar).^{11,12} To prevent adverse sequelae, this technique may be tested on an easily concealed area prior to more aggressive treatment. Compared to CO₂ laser vaporization, low-current needle tip cautery may provide equal or superior cosmesis. The rate of recurrence may be lower due to the deeper skin penetration obtained with electrocautery as compared to the more superficial penetration of an ablative CO₂ laser. The laser also does not permit tissue to be sent for histopathologic examination.

In summary, the technique described here in a large series of patients represents a novel method of removing numerous lesions in patients with neurofibromatosis. The ease of performing the excision in an ambulatory setting, the speed with which it can be accomplished, the minimal discomfort it causes, the excellent aesthetic results and the overall patient satisfaction are reasons

why this may be the optimum method for removal of cutaneous neurofibromas.

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