Successful radical surgical resection of a giant neurofibroma

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Abstract

Large neurofibromas often cause significant patient morbidity and present a unique challenge to dermatologists and surgeons. Radical resection offers the lowest rate of recurrence but is not often pursued due to the high risk of intraoperative hemorrhage and difficulty in repairing large defects. Subtotal resection and debulking are more frequently performed, leading to higher rates of recurrence. This case highlights a particularly large neurofibroma and demonstrates how surgical techniques like preoperative embolization and advancement flaps can improve outcomes in the radical resection of large neurofibromas.

Keywords: advancement, embolization, flap, neurofibroma, neurofibromatosis, surgery

Introduction

Neurofibromatosis type one (NF1) is a relatively rare (estimated birth incidence of one in 2500) autosomal dominant genetic condition caused by mutations of the *NF1* gene and characterized by multiple skin alterations such as café-au-lait macules, axillary freckling, and neurofibromas [1]. Neurofibromas are benign peripheral nerve sheath tumors composed of Schwann cells, perineurial-like cells, and fibroblasts [2]. Although benign, large neurofibromas often result in significant patient morbidity and present a unique challenge to dermatologists and surgeons. Radical resection offers the lowest rate of recurrence but is not often pursued due to the high risk of intraoperative hemorrhage and difficulty in repairing large defects. Subtotal resection and debulking are more frequently performed, leading to higher rates of recurrence [3].

Case Synopsis

A 32-year-old man with a history of NF1 presented with an enlarging growth along his right lateral neck and supraclavicular region. The growth started in childhood as a relatively flat, velvety plaque. A localized resection was performed when the patient was 12 years old, but it regrew and doubled in size over the past three years. Clinical examination revealed a 22×44cm hyperpigmented, firm pedunculated mass on the right lateral neck and supraclavicular region suspended over the chest (**Figure 1A**). The large size required the patient to use a specialized brace to hold the mass in place in order to use his right hand. Surgical intervention was recommended as continued growth was likely to threaten his airway.

Positron emission tomography was notable for a large hypermetabolic exophytic soft tissue mass

centered in the right supraclavicular region. Magnetic resonance imaging confirmed the presence of a mass suggestive of neurofibroma with abundant vascular flow.

The patient underwent embolization with interventional radiology, followed by resection and repair with an advancement flap by plastic and thoracic surgery. The wound was completely healed by post-operative day 42 (**Figure 1B**). Surgical pathology revealed a loose spindle cell neoplasm comprised of cells with ovoid nuclei set in a delicate collagenous background with prominent blood vessels and entrapped adnexal structures, diagnostic of a diffuse cutaneous neurofibroma (**Figure 1C**).

Case Discussion

Neurofibromas are categorized based on their appearance, anatomic location, and histopathology as cutaneous (localized or diffuse), intraneural, plexiform, or massive soft tissue neurofibromas. Localized cutaneous neurofibromas are the most common form and grows as a nodular and well-circumscribed lesion of the skin. Diffuse cutaneous neurofibromas feature more extensive involvement of the skin and subcutaneous tissue, commonly affecting small nerves. Intraneural neurofibromas grow within nerve fibers and cause fusiform enlargement of affected nerves. Plexiform neurofibromas are multinodular elongated masses that most often affect large nerves. Massive soft tissue neurofibromas causes diffuse infiltration of the soft tissue of a specific area, usually the distal extremity [2].

Histopathology can help differentiate between diffuse cutaneous and plexiform subtypes, as both can be large and commonly occur around the head and neck [4]. Microscopically, cutaneous neurofibromas feature expansile proliferation around normal structures, such as skin, adnexa, and blood vessels. Plexiform neurofibromas commonly form a tangled mass of tumors, each representing a nerve or nerve fascicle distended by tumor cells embedded in a rich myxoid matrix [5]. In this case, tumor infiltration within blood vessels and adnexal structures was diagnostic of diffuse cutaneous neurofibroma.

Conclusion

This case provides an example of a diffuse cutaneous neurofibroma that was particularly large and debilitating for the patient. It highlights how surgical techniques like preoperative embolization and advancement flaps can improve outcomes in the radical resection of large neurofibromas.

Potential conflicts of interest

The authors declare no conflicts of interest.

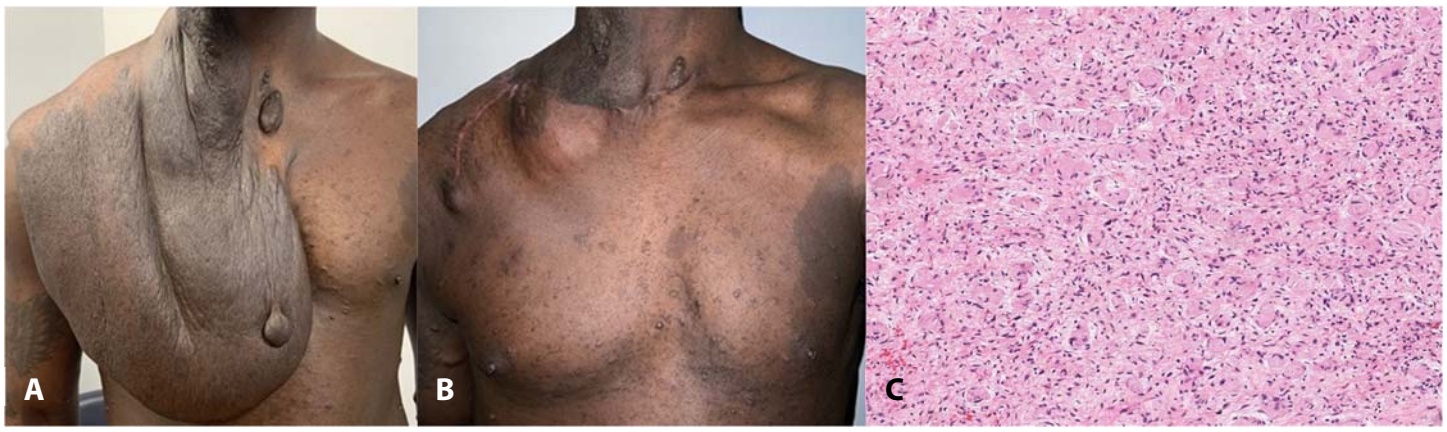

Figure 1. ***A)*** *The 22×44cm fleshy hyperpigmented pedunculated mass originating from the right lateral neck and supraclavicular region.* ***B)*** *Postoperative outcome with a well-healed scar along the right supraclavicular region.* ***C)*** *The excised specimen demonstrates wavy spindled cells in a delicate collagenous background with entrapped adnexal structures. Increased cellularity without significant nuclear atypia or mitotic activity is noted. H&E, 100×.*

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