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Insights of Clinical Case Reports

Case Report

Immediate Nipple Areolar Complex Reconstruction in a Patient with Von Recklinghausen's Neurofibromatosis: A Clinical Case Report

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1. Abstract

Von Recklinghausen's neurofibromatosis is an autosomal dominant condition commonly characterized by café au lait patches, axillary freckling, multiple neurofibromas, and Lisch nodules. In this context, we present a complex case involving a 32-year-old female who suffered from severe neurofibromatosis, leading to a significant impact on her bilateral nipple-areolar complexes. Furthermore, we illuminate the subsequent outcome of her immediate reconstruction procedure.

2. Keywords: Breast ; Neurofibromatosis ; Nipple ; Plastic Surgery

3. Introduction

Neurofibromatosis type 1 [NF1] is an autosomal dominant disorder, with an estimated incidence ranging from 1 in 2,500 to 1 in 3,000 [1], of which 50% of cases are sporadic [2]. Individuals with NF1 exhibit diverse manifestations, including café au lait spots, neurofibromas, intertriginous freckling, Lisch nodules, and bony dysplasia in long bones, all stemming from the deficiency of neurofibromin protein.

Neurofibromas in the breast are uncommun [3–5], particularly those affecting the nipple-areolar complex. This paper aims to present a case of a female patient with neurofibromas involving the nipple-areolar complex, while also discussing the surgical approach and the optimal timing for reconstruction.

4. Clinical Case

A 32-year-old woman diagnosed with NF1 based on the National Institutes of Health criteria [6] was referred to our department due to the significant and deforming neurofibromatosis affecting both bilateral nipple-areolar complexes (Figure 1). Notably, there was no family history of NF1. Upon examination, numerous cutaneous neurofibromas were evident across her chest and breasts, with particularly pronounced serpiginous, pedunculated neurofibromas on both sides. These lesions caused pain and protruded outward by 3 cm to 5 cm from the nipple and areola, resulting in deformity of both nipples. Over a span of two years, these neurofibromas had progressively increased in size. The patient was significantly distressed by the disfigurement caused by these neurofibromas and actively sought their removal.Mammography revealed well-defined, dense, pedunculated nodules originating from the areolar region. In light of the clinical presentation, the decision was made to proceed with localized excision of the neurofibromas, coupled with minor plastic surgery interventions to restore symmetry to the areolar region. The nipples were reshaped using a CV flap technique, and the areola was reconstructed with a total skin graft (Figure 2). Post-operatively, the patient experienced no complications and expressed satisfaction with the final aesthetic outcome. The histopathological examination verified the neural differentiation of spindle cells and the presence of Meissner corpuscles, demonstrating immunopositivity for S100. However, proliferation was observed to be negative for epithelial membrane antigen [EMA].

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Received date: Dec 22, 2023; **Accepted date:** Jan 11, 2024; **Published date:** Jan 16, 2024

Citation: Ghorbel I (2024). Immediate Nipple Areolar Complex Reconstruction in a Patient with Von Recklinghausen's Neurofibromatosis: A Clinical Case Report. Insig of Clin and Med Case Rep 2024; v1(1): 1-3

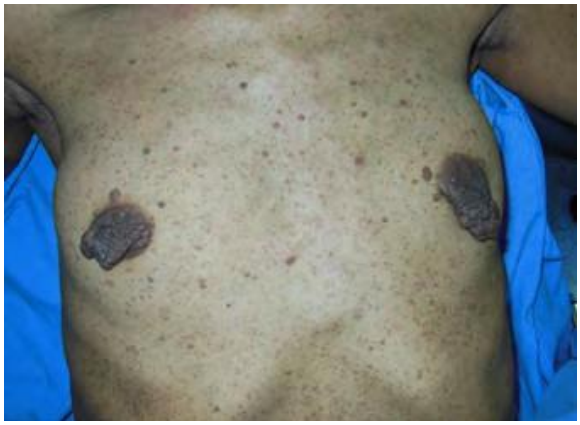

Figure 1: Neurofibromatosis involving bilateral nipple areolar complex

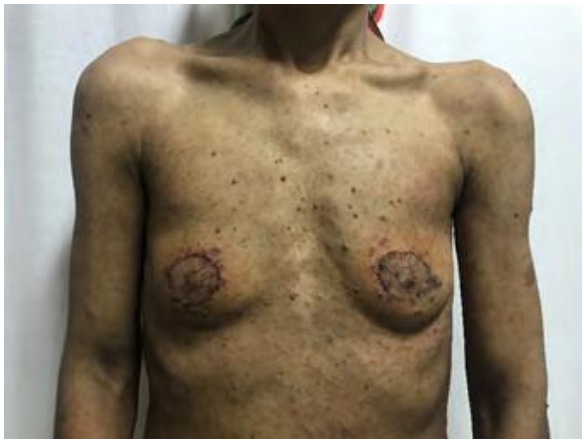

Figure 2: Post local excision of neurofibromas + Reconstruction of nipple areolar complex

5. Discussion

Neurofibromas occurring in the breast are exceedingly uncommon [3,4]. Only a limited number of reported cases have discussed treatment approaches, especially when these neurofibromas involve the nipple-areolar complex [5]. A study conducted by Freidrich et al, uncovered that tumor emerging in the areola and nipple region of NF1 patients, which resembled accessory nipples, were all identified as neurofibromas. This phenomenon was observed in both males and females, with occasional occurrences in children, showing a greater inclination for females to develop cutaneous neurofibromas in this particular location [3].

In general, neurofibromas tend to be asymptomatic; surgical intervention may be considered for cosmetic reasons when tumors reach a substantial size [7]. When these neurofibromas grow significantly and become unwieldy, they are characterized using terms such as “elephantiasis neuromatosa” or “fibroma pendulum” [4].

Therapeutic options include surgical excision for managing painful and disfiguring lesions, as well as treatment using CO2 laser [8]. Previous reviews have showcased instances where surgical intervention was limited to localized removal of neurofibromas from the nipple area, involving procedures like nipple and areola reduction [3,4,9]. To our knowledge, this case report stands as the first documentation of immediate nipple reconstruction using a CV flap following the excision of neurofibromatous lesions affecting the nipple-areolar complex. We performed a thorough excision of both nipple-areolar complexes, managing the resultant skin defect by utilizing purse-string sutures. Simultaneously, within the same surgical session, we employed a CV technique to create the nipple and implemented a skin graft to reconstruct and restore the integrity of the areolar region.

Our patient reported a high level of satisfaction following the surgery.

In cases similar cases, complete excision of neurofibromas coupled with reconstruction not only ensures favorable aesthetic outcomes [10] but also serves as a safeguard against potential sarcomatous transformation [11].

It’s important to acknowledge that NF1 patients face an elevated risk of developing various benign and malignant neoplasms, including breast cancer [12–14].

There has been conjecture surrounding the possible effect of numerous neurofibromas within the breast and the nipple-areolar complex, which might inadvertently obscure the detection of a breast mass during palpation, potentially leading to delayed clinical recognition [15–17]. In cases where a breast lesion is suspected in patients with NF1, it is recommended to undergo radiological imaging to gather additional diagnostic insights [7,18].

The breast holds significant importance within the framework of female identity. We firmly assert that preserving the nipple-areolar complex [NAC] is of utmost importance in the process of restoring a patient’s body image [10,19]. Moreover, individuals with NF1, including our patient, frequently encounter discrimination or rejection from society, and even within their own families. This can be attributed to the distressing appearance associated with NF1, as well as the potential presence of learning and behavioral disabilities that often accompany the condition [2,17]. In such cases, plastic surgery not only addresses the physical aspects but also offers a chance for these individuals to regain a sense of normalcy and social acceptance.

6. Conclusion

Our case of bilateral neurofibroma excision, which included the nipple-areolar complex, followed by immediate reconstruction in a female diagnosed with NF1, remains an unusual occurrence in clinical practice.

The significance of plastic surgery in the treatment of breast neurofibromas among NF1 patients cannot be overstated, as it plays a crucial role in preserving function and psychological equilibrium.

We advocate for an early and meticulous physical assessment, alongside the utilization of various imaging techniques for screening purposes, whenever any potentially concerning clinical indicators manifest in patients with NF1.

7. Author Contributions

All authors contributed significantly and were in agreement with the content of the manuscript. All authors participated in data collection and in writing the manuscript.

8. Conflicts of Interest

No author has any financial or personal relations- hip with any person or organization that could inappropriately influence their work.

References

1. Gutmann DH, Ferner RE, Listernick RH, Korf BR, Wolters PL, Johnson KJ. Neurofibromatosis type 1. Nat Rev Dis Primers. 2017; 3: 17004.
2. Ly KI, Blakeley JO. The Diagnosis and Management of Neurofibromatosis Type 1. Med Clin North Am. 2019; 103(6): 1035-1054.
3. Friedrich RE, Hagel C. Appendices of the nipple and areola of the breast in Neurofibromatosis type 1 patients are neurofibromas. Anticancer Res. 2010; 30(5): 1815-7.
4. Hero E, Carey M, Hero I, Shaaban AM. Bilateral Neurofibromas of the Nipple-Areolar Complex: A Case Report and Approach to Diagnosis. Case Rep Pathol. 2018; 2018: 6702561.
5. Lannon DA, Harris PA, Butler PEM. Treatment of severe neurofibromatosis of the nipple-areola complex. Plast Reconstr Surg. févr 2003; 111(2): 952-3.
6. Neurofibromatosis. Conference statement. National Institutes of Health Consensus Development Conference. Arch Neurol. Mai. 1988; 45(5): 575-8.
7. Aljindan F, Aljehani L, Alsharif B, Mortada H. Management of Neurofibromatosis of the Nipple-Areolar Complex. Case Rep Surg. 2021; 2021: 6622416.
8. Ostertag JU, Theunissen CCW, Neumann HAM. Hypertrophic scars after therapy with CO2 laser for treatment of multiple cutaneous neurofibromas. Dermatol Surg Off Publ Am Soc Dermatol Surg Al. mars 2002; 28(3): 296-8.
9. White CP, Martin S, Dal Cin A. Surgical treatment of areolar neurofibromas. Eur J Plast Surg. Août. 2012; 35(8): 625-7.
10. Satteson ES, Brown BJ, Nahabedian MY. Nipple-areolar complex reconstruction and patient satisfaction: a systematic review and meta-analysis. Gland Surg. Févr. 2017; 6(1): 4-13.
11. Uddin W, Ahmad F, Khawar A. Recurrent neurofibroma in the head and neck. J Coll Physicians Surg Pak. 2007; 17(10): 629-31.
12. Seo YN, Park YM. Association between Neurofibromatosis Type 1 and Breast Cancer: A Report of Two Cases with a Review of the Literature. Case Rep Med. 2015; 2015: 456205.
13. Seminog OO, Goldacre MJ. Age-specific risk of breast cancer in women with neurofibromatosis type 1. Br J Cancer. 28 avr 2015; 112(9): 1546-8.
14. Da Silva AV, Rodrigues FR, Pureza M, Lopes VGS, Cunha KS, et al. Breast cancer and neurofibromatosis type 1: a diagnostic challenge in patients with a high number of neurofibromas. BMC Cancer. 26 mars 2015; 15: 183.
15. Bongiorno MR, Doukaki S, Aricò M. Neurofibromatosis of the nipple-areolar area: a case series. J Med Case Reports. 2010; 4: 22.
16. Liu T, Liang Y, Lalwani SK, Zhang Y. Paget disease of the breast with underlying invasive ductal cancer in neurofibromatosis type 1: a case report. Transl Cancer Res. 2023; 12(3): 638-645.
17. Zhou Y, Pan B, Mao F, Zhu Q, Huo Z, Wang H, et al. A hidden breast lump covered by nipple appendices in a patient with von recklinghausen disease: a case report and review of the literature. Clin Breast Cancer. févr 2012; 12(1): 71-5.
18. Nasri S, Benmoussa Y, Abbou W, Mirali H, Aichouni N, Skiker I, et al. Imaging appearance of isolated diffuse neurofibroma of nipple areolar area: a case report. Pan Afr Med J. 2021; 39: 178.
19. Lee JS, Eom JR, Lee JW, Choi KY, Chung HY, Cho BC, et al. Safe delayed procedure of nipple reconstruction in poorly circulated nipple. Breast J. janv. 2019; 25(1): 129-33.