Surgical resection of a solitary neurofibroma. A case report

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Background: Neurofibromatosis type 1 (NF1) is an autosomal dominant RASopathy caused by pathogenic variants in the NF1 gene, with a pooled global prevalence of approximately one in 3,164 individuals. Cutaneous and plexiform neurofibromas are hallmark features of the disease; although most cutaneous lesions follow an indolent course, those arising in cosmetically and functionally sensitive regions such as the nose pose distinct diagnostic and surgical challenges, particularly in pediatric patients. We report the case of a 14-year-old female with a clinical diagnosis of NF1 who presented with a neurofibroma involving the internal portion of the left nasal ala. The patient had been started on selumetinib, a MEK1/2 inhibitor used for symptomatic plexiform neurofibromas, and subsequently developed an eczematous cutaneous reaction that prompted evaluation by the allergy and immunology service. She was admitted to the reconstructive surgery service for elective resection of the nasal lesion under a multidisciplinary approach involving pediatric neurology, plastic surgery, and allergy/immunology. This report describes the clinical presentation, multidisciplinary decision-making, and surgical management of the case, and discusses the dermatologic adverse effects of MEK-inhibitor therapy that can influence the timing and coordination of surgery in pediatric NF1 patients. To our knowledge, few reports address the concurrent surgical and pharmacologic management of a resectable nasal neurofibroma during ongoing selumetinib therapy, underscoring the educational relevance of this case.

Keywords: Neurofibromatosis type 1, nasal neurofibroma, selumetinib, MEK inhibitor, pediatric plastic surgery, RASopathy, cutaneous neurofibroma, case report

Neurofibromatosis type 1 (NF1), formerly known as von Recklinghausen disease, is one of the most common autosomal dominant genetic disorders, with a birth incidence estimated at one in 2,662 and a pooled prevalence of one in 3,164 individuals worldwide [1]. It results from pathogenic variants in the NF1 gene on chromosome 17q11.2, which encodes neurofibromin, a GTPase-activating protein that negatively regulates the RAS-MAPK signaling pathway; loss of neurofibromin function leads to constitutive RAS activation and a predisposition to both benign and malignant tumors of neural crest origin [2]. The clinical diagnosis of NF1 has historically relied on the 1987/1997 NIH consensus criteria, which were revised in 2021 by an international consensus panel to incorporate genetic testing and additional clinical features, shortening the time to diagnosis in pediatric cohorts [3,4].

Neurofibromas, the clinical hallmark of NF1, are classified as cutaneous (superficial, arising from small dermal nerve twigs) or plexiform (deeper, congenital, involving multiple nerve fascicles) [5]. While cutaneous neurofibromas are almost universally benign, plexiform neurofibromas carry an estimated 8-13% lifetime risk of malignant transformation into a malignant peripheral nerve sheath tumor (MPNST) [5,6]. Facial and nasal involvement, although less common than trunk or limb disease, has a disproportionate impact on quality of life: visible craniofacial neurofibromas are associated with reduced self-esteem, anxiety, and increased vulnerability to bullying in children and adolescents with NF1 [7]. Isolated neurofibromas of the nasal ala or nasal tip are rare in the pediatric literature and typically warrant surgical excision to preserve nasal aesthetic subunits and function while minimizing recurrence [8,9].

Surgical management remains the mainstay of treatment for accessible, symptomatic, or cosmetically significant neurofibromas [10,11]. For diffuse or inoperable plexiform disease, selumetinib, a selective MEK1/2 inhibitor, was approved in 2020 for pediatric patients two years of age and older with symptomatic, inoperable plexiform neurofibromas, after demonstrating durable tumor volume reduction and improvement in patient-reported morbidity in the SPRINT trial [12,13]. Dermatologic adverse effects—including acneiform and maculopapular rash, xerosis, paronychia, and less commonly eczematous reactions—are among the most frequent toxicities

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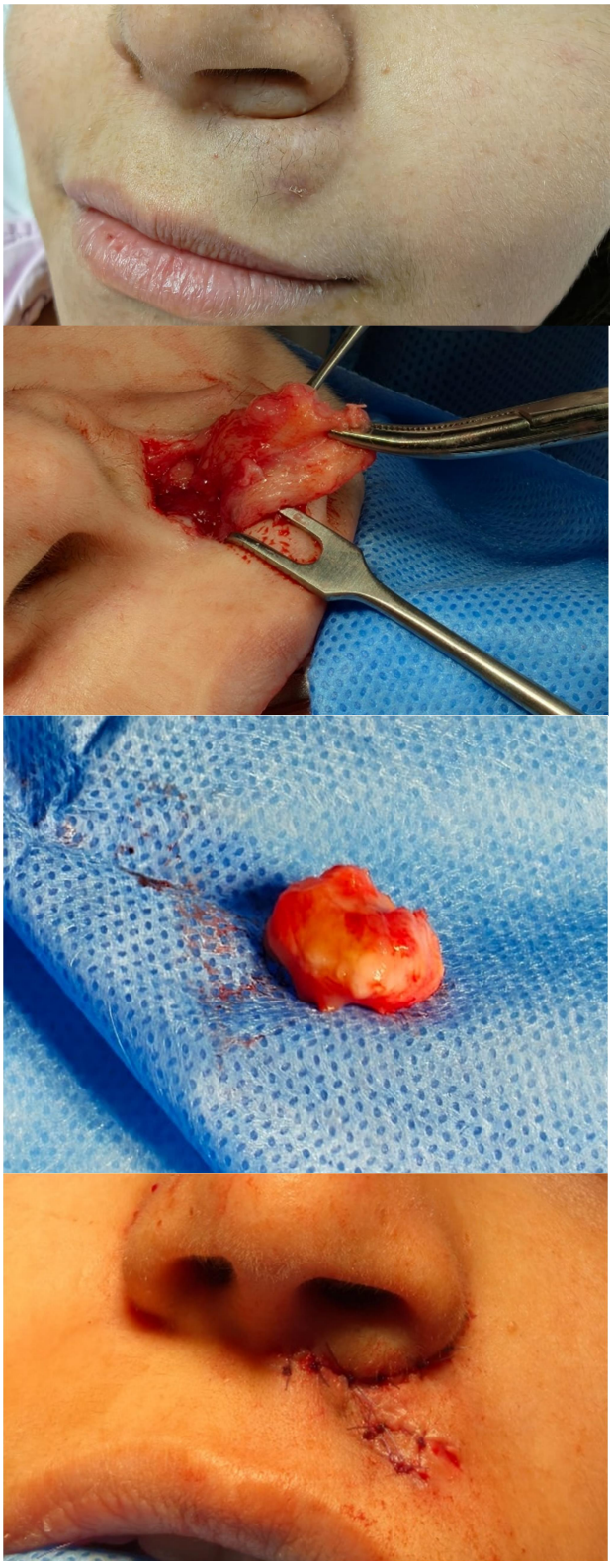

associated with selumetinib and can require dose adjustment or dermatologic/allergy co-management [14-16]. We present the case of an adolescent with NF1 and a solitary neurofibroma of the left internal nasal ala who underwent surgical resection while concurrently receiving selumetinib for her broader neurofibromatous disease, complicated by an eczematous drug reaction. This case was prepared following the CARE (CAse REport) guidelines for case report reporting [17].

Case report

A 14-year-old female with a clinical diagnosis of neurofibromatosis type 1, followed by the pediatric neurology service, presented with a neurofibroma involving the internal portion of the left nasal ala. Which as referred by the patient, presented numbness, andc aused progressive disconmfort for activities involving the mouth, such as eating, washing teeths or drinking. (Figure 1)

Family history was notable for a mother with systemic arterial hypertension and hypothyroidism, a healthy father, a maternal grandmother with diabetes mellitus, a maternal grandfather with systemic arterial hypertension, and a paternal grandmother with type 2 diabetes and hypertension; no family history of neurofibromatosis was documented, consistent with either a de novo NF1 variant or an incompletely characterized family history. Personal history was unremarkable: no known allergies, chronic diseases, prior fractures, hospitalizations, surgeries, or transfusions, and a complete vaccination schedule for age.

As part of her neurofibromatous disease management, the patient had been started on selumetinib. She subsequently developed an eczematous cutaneous reaction, prompting an urgent interconsultation to the allergy and immunology service for evaluation and co-management, coordinated with the pediatric neurology team.

On admission to the reconstructive surgery service, vital signs were: blood pressure 111/70 mmHg, heart rate 80 beats per minute, temperature 37.6°C, respiratory rate 20 breaths per minute, and oxygen saturation 96% on room air; weight was 46 kg and height 156 cm. Physical examination was notable for a neurofibroma involving the internal portion of the left nasal ala, with the left nasal fossa otherwise normochromic and turbinates in cycle; the right nasal fossa was normochromic without lesions. Otoscopy was bilaterally unremarkable, the oropharynx was without hyperemia, exudate, or postnasal discharge, and no cervical adenopathy was identified. Cardiopulmonary, abdominal, and extremity examinations were within normal limits, and no other cutaneous, skeletal, or neurologic abnormalities were reported on the systems review.

The patient was admitted under the reconstructive/plastic surgery service for elective surgical resection of the nasal neurofibroma. A preoperative surgical safety checklist was completed per institutional protocol, confirming patient identity,

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procedure, and anesthetic safety measures. The very next day, the patient underwent surgical excision of the neurofibroma of the internal portion of the left nasal ala. After the recovery from anethesia, and with the next vital signs acquired blood pressure 100/64 mmHg, Heart Rate 74, Temperature 36.8°C, Respiratory Rate 18 bpm, and Oxygen Saturation 94% with out the use of supplementary oxygen, the patient was discharged and cited for the follow up checkups the veyr next week.

Discussion

This case illustrates two intersecting aspects of NF1 management in pediatric patients: the surgical approach to a localized, resectable neurofibroma in a cosmetically sensitive facial region, and the coordination required when a patient is concurrently receiving systemic MEK-inhibitor therapy for more extensive neurofibromatous disease. NF1 has nearly complete penetrance by adulthood but a highly variable phenotype, and craniofacial involvement—although present in a minority of patients—carries disproportionate psychosocial weight, given its visibility and its association with reduced self-esteem and peer victimization in adolescents [7,18]. Isolated, well-circumscribed neurofibromas of the nose, such as reported in an 11-year-old girl with a nasal tip lesion and in a 10-year-old boy with midface plexiform involvement causing nasal and labial asymmetry, are amenable to surgical excision with generally favorable cosmetic and functional outcomes, provided the aesthetic subunits of the nose are respected during planning [8,9].

Surgical treatment continues to be considered first-line therapy for accessible NF1-associated neurofibromas, including plexiform lesions amenable to complete or near-complete resection [10,11]. However, for patients with diffuse, deep, or otherwise inoperable plexiform disease causing significant morbidity, selumetinib has become an important pharmacologic option, having demonstrated volumetric tumor reduction and functional improvement in the pivotal SPRINT trial that supported its 2020 FDA approval for patients two years of age and older [12,13]. Dermatologic toxicity is among the most frequently reported adverse effects of selumetinib, encompassing acneiform and maculopapular rash, xerosis cutis, paronychia, and, less commonly, eczematous reactions, as described in consensus management recommendations and in real-world pediatric cohorts [14-16]. In our patient, the eczematous reaction required prompt allergy/immunology co-management, highlighting the importance of close dermatologic surveillance and multidisciplinary coordination between medical oncology/neurology, allergy, and surgical teams when a patient is receiving concurrent pharmacologic and surgical treatment for NF1-associated tumors.

From a surgical standpoint, plastic and reconstructive surgery plays a central role across the clinical spectrum of NF1-associated nerve sheath tumors, from small cutaneous and subcutaneous lesions to large, deforming plexiform disease, with the timing and extent of surgery individualized according to symptoms, functional and aesthetic impact, disease progression, and malignant potential [11]. Although cutaneous neurofibromas carry a low risk of malignant transformation, plexiform neurofibromas are associated with an estimated lifetime MPNST risk of 8-13%, underscoring the value of histopathologic confirmation after excision and of longitudinal surveillance for red-flag features such as rapid growth, new pain, or neurologic deficit [5,6]. This case is limited by the absence, at the time of this draft, of definitive histopathology and postoperative follow-up data, which will be incorporated once available; nonetheless, it adds to the limited body of literature describing concurrent surgical and MEK-inhibitor management of NF1 in the pediatric nasal region.

Conclusion

Neurofibromas of the internal nasal ala are an uncommon but clinically significant manifestation of neurofibromatosis type 1 in pediatric patients, and their surgical resection requires careful preservation of nasal aesthetic and functional subunits. When surgery is undertaken in a patient concurrently receiving selumetinib for broader neurofibromatous disease, close multidisciplinary coordination between plastic surgery, pediatric neurology, and allergy/immunology is essential to manage treatment-related dermatologic toxicity without delaying indicated surgical care. Reporting the surgical and pharmacologic course of such cases in detail can help guide clinicians managing similar overlapping scenarios.

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