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Management of multiple facial leiomyomas with excision and cervicofacial flap reconstruction in a severe Hemophilia A patient: A case presentation

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The authors stated that the written consent was obtained from the patient presented with images in the study.

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Abstract

Cutaneous leiomyomas are rare, benign, smooth-muscle tumors that are particularly uncommon on the face. Surgical management of cutaneous leiomyomas in patients with severe Hemophilia A is challenging because of the substantial risk of bleeding, particularly when regional flap reconstruction is required. Here, we report the case of a 64-year-old male with severe Hemophilia A who presented with multiple painful cutaneous nodules in the right preauricular and submandibular region. MRI and biopsy confirmed multiple cutaneous leiomyomas of piloleiomyoma and angioleiomyoma types. A multidisciplinary team developed a perioperative recombinant factor VIII protocol to minimize bleeding risk. Surgical excision with a narrow 5 mm margin was performed, followed by cervicofacial flap reconstruction. The patient's intra- and immediate postoperative course was uneventful. Flap viability was maintained despite minor marginal ischemia, which resolved with conservative management. Factor VIII levels remained within therapeutic range throughout hospitalization, and histopathology confirmed the leiomyomas' benign status. Renal cancer screening was recommended because of the association with multiple leiomyomas. The patient achieved satisfactory aesthetic and functional recovery without bleeding complications. This case highlights that complex facial excision and flap reconstruction can be performed safely in patients with severe Hemophilia A using a tailored factor VIII replacement protocol and underscores the importance of multidisciplinary perioperative planning in such cases.

Keywords: hemophilia A, cervicofacial flap reconstruction, cutaneous leiomyoma, factor VIII replacement protocol, hemostasis

Introduction

Cutaneous leiomyomas are benign, smooth-muscle tumors that represent approximately 5% of all benign soft-tissue tumors [1]. Facial presentation is particularly rare, with fewer than 100 cases reported in the literature [2]. When these tumors occur in multiple locations, they may be associated with Reed's Syndrome (Multiple Cutaneous and Uterine Leiomyomatosis), which requires additional screening for renal cell carcinoma.

Severe Hemophilia A presents significant challenges for surgical intervention, with reported perioperative bleeding rates as high as 32% in major procedures [3].While factor replacement guidelines exist for orthopedic and abdominal surgeries, limited protocols are available for large facial excisions with a regional flap reconstruction [4]. This report addresses this gap and outlines a multidisciplinary approach to the successful management of multiple cutaneous leiomyomas of the face in a patient with severe Hemophilia A.

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Case presentation

Patient Information

A 64-year-old male with severe Hemophilia A presented with multiple painful nodular lesions localized to the right preauricular, submandibular, and upper cervical region. The patient reported pain exacerbated with cold exposure and aesthetic concerns. His past medical history included treated Hepatitis C and hypertension. There was no family history of similar lesions or syndromic features, and no preoperative genetic testing was performed for Reed's Syndrome or fumarate hydratase mutations.

Clinical Findings

Physical examination revealed multiple firm, tender, erythematous nodules over the right preauricular and submandibular area that extended posteriorly from the posterior mandibular angle to the anterior mandibular body, superiorly from the mid tragus to the hyoid bone (Figure 1). The largest lesion measured 2.5 cm in diameter.

Diagnostic Assessment

MRI revealed multiple cutaneous lesions in the submandibular and upper cervical region, isointense on T1-weighted imaging and mildly hyperintense on T2, with homogeneous contrast enhancement. No deep extension or pathological lymphadenopathy was observed. The imaging suggested a benign status; however, due to the multifocal nature and size of the lesions, malignancy could not be entirely excluded. Surgical biopsy was performed that revealed multiple cutaneous leiomyomas of piloleiomyoma and angioleiomyoma types.

Factor VIII Protocol

A comprehensive factor VIII protocol was implemented in collaboration with hematology. Immediately prior to incision, 3,000 units of recombinant factor VIII and 1 g of tranexamic acid were administered. Twelve hours postoperatively, 1,000 units of factor VIII were administered. On postoperative days 1–3, 2,000 units were administered each morning and 1,000 units were administered each evening, with factor VIII activity and aPTT measured on postoperative days 2 and 3. On postoperative days 4–7, 2,000 units were administered daily; on days 8, 10, 12 and 14, 3,000 units were administered each morning. Tranexamic acid was continued orally until day 10. No inhibitors were detected, and hemostasis was maintained without additional agents. Factor VIII activity remained between 21% and 36 % during the first postoperative week, which guided ongoing supplementation to ensure effective hemostasis.

Therapeutic Intervention

Under general anesthesia without muscle relaxants, a local excision with a narrow 5 mm margin was performed (Figure 2). The surgical specimen measured 10.5×8.0×0.5 cm. A cervicofacial flap reconstruction was performed with preservation of the external jugular vein and the greater auricular nerve (Figure 3). The flap was raised deep to the platysma (Figure 3) and was rotated and advanced to cover the defect. Closure was achieved using Vicryl 3-0 and 4-0 sutures for subcutaneous layers and Nylon 3-0 and 4-0 for the skin. A surgical drain was placed.

Pathological Findings

Histopathology revealed multiple cutaneous piloleiomyoma and angioleiomyoma types. The specimen contained multiple nodules, with the largest measuring 1.5 cm in diameter. A clinical workup for renal neoplasms was recommended due to the multiplicity of the lesions.

Figure 1. Preoperative clinical appearance showing multiple nodular lesions in the right preauricular and submandibular regions.

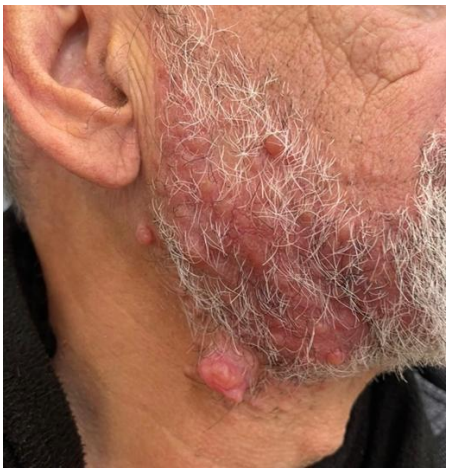


Figure 2. Intraoperative view of local excision of the cutaneous leiomyomas with narrow margins.

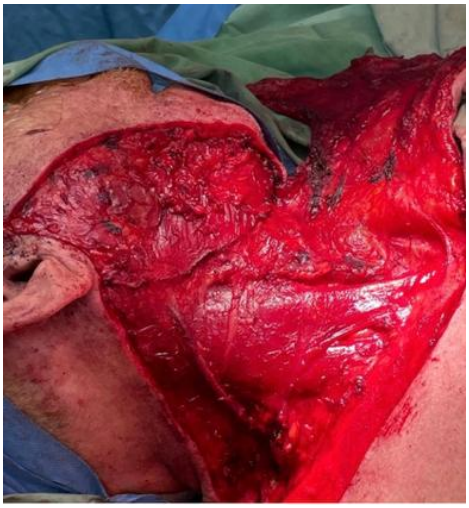
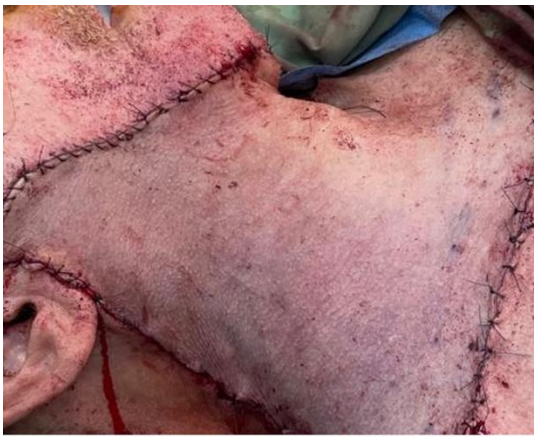


Figure 3. Elevation of the cervicofacial flap deep to the platysma for defect reconstruction.



Follow-up and Outcomes

The patient had an uncomplicated initial recovery. He remained afebrile with stable vital signs, and facial nerve function was preserved. On postoperative day 3, discoloration of the distal flap edge raised concern for marginal ischemia. The area was managed conservatively with daily flap stimulation and dressings. A small area of necrosis demarcated and was scheduled for outpatient debridement. The patient was treated with daily Vaseline dressings until granulation tissue epithelialized. The patient achieved satisfactory aesthetic and functional outcomes (Figure 4 and Figure 5), with resolution of temperature-related pain. No further complications were noted during follow-up.

Figure 4. Immediate postoperative result following cervicofacial flap inset and closure.

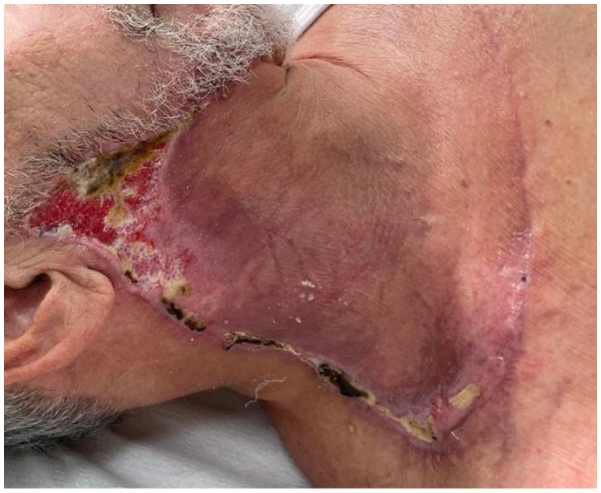
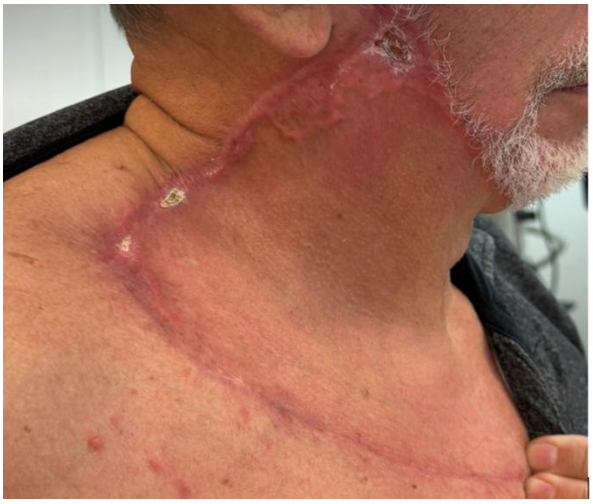


Figure 5. Healed postoperative outcome showing satisfactory aesthetic and functional results.



Discussion

This case demonstrates the feasibility of major head-and-neck reconstruction in cases of severe hemophilia when perioperative hemostasis is managed meticulously. The factor VIII protocol used here builds upon established recommendations but was tailored for head-and-neck surgery, where rich vascularity increases bleeding risk [5].

Importantly, the cervicofacial flap offered broad coverage, excellent color and texture match, and minimized the creation of new surgical planes, reducing potential bleeding surfaces. Alternative options such as submental or nasolabial flaps were considered but would have entailed additional donor-site morbidity and higher bleeding risk. To the best of our knowledge, the use of a cervicofacial flap in a patient with severe hemophilia has not been previously reported.

The coexistence of piloleiomyoma and angioleiomyoma components triggered consideration of Reed's Syndrome. However, this syndrome typically occurs in women with uterine leiomyomas. Because multiple cutaneous leiomyomas can indicate a hereditary tumor syndrome, renal-cell-carcinoma screening was recommended [2]. Overall, this successful outcome underscores the critical role of multidisciplinary planning, close hematologic collaboration, careful surgical technique, and vigilant postoperative monitoring in cases of cutaneous leiomyomas in patients with severe Hemophilia A.

Conclusion

This case demonstrates that complex facial reconstruction can be safely performed in severe hemophilia patients with appropriate perioperative planning. The detailed factor VIII protocol presented here may serve as a guide for similar cases. The use of a cervicofacial flap for reconstructive surgery in this context is a novel approach, highlighting the importance of interdisciplinary collaboration. This report underscores the need for future studies to develop guidelines for facial surgery in hemophilia patients.

References

1. Dhankar N, Tomar R, Paul S, Khurana N, Neogi S. Leiomyoma of the nipple: a rare entity at a rare site. Cureus2024;16:e62220. doi:10.7759/CUREUS.62220
2. Winchester DS, Hocker TL, Brewer JD, Baum CL, Hochwalt PC, Arpey CJ, et al. Leiomyosarcoma of the skin: clinical, histopathologic, and prognostic factors that influence outcomes. J Am Acad Dermatol 2014;71:919–25. doi:10.1016/j.jaad.2014.07.020
3. Suzan V, Ozgur Yurttas N, Eskazan AE, Ozmen D, Keskin D, Sadri S, et al. Management of major surgical procedures in patients with hemophilia: a single-center experience of 49 procedures. Blood 2019;134:4933-3. doi:10.1182/BLOOD-2019-130305
4. Poston JN, Kruse-Jarres R. Perioperative hemostasis for patients with hemophilia. Hematology 2022;2022:586-93. doi:10.1182/HEMATOLOGY.2022000387
5. Srivastava A, Santagostino E, Dougall A, Kitchen S, Sutherland M, Pipe SW, et al. WFH Guidelines for the Management of Hemophilia, 3rd edition. Haemophilia 2020;26(Suppl 6):1-158. doi:10.1111/HAE.14046