

Squamous Carcinoma of the Oral Cavity in Patient with Fanconi Anemia: A Challenging Reconstruction with Double-barrel Free Fibula Flap Case Report

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Summary: Fanconi anemia (FA) is a very rare form of aplastic anemia. Patients with FA have a higher risk of developing solid tumors such as head and neck squamous cell carcinoma, higher risk of local recurrence, and impaired resistance to chemotherapy and radiotherapy treatments than the normal population. In this article, we describe the challenging clinical case of a patient with FA who underwent surgery for the removal of a large squamous cell carcinoma in the oral cavity. Mandibular reconstruction was performed using a biaxial double-barrel fibular flap, with excellent functional aesthetic outcomes. (*Plast Reconstr Surg Glob Open* 2023; 11:e5414; doi: 10.1097/GOX.0000000000005414; Published online 20 November 2023.)

In 1927, Guido Fanconi's description of a familial aplastic anemia form in three brothers marked the inception of Fanconi anemia (FA).¹ Subsequently, it has evolved into a syndrome encompassing bone marrow hypoplasia, skeletal anomalies, renal impairments, ophthalmological deformities, and chromosomal aberrations. FA's occurrence in the general population is one to five cases per million, with nearly half of Italy's FA instances being clustered in Campania. Underlying FA is the loss of function of genes involved in the DNA repair pathways. As a result, patients with FA face heightened risks of solid tumors and leukemia, even at a young age.² Head and neck squamous cell carcinoma is the most common solid tumor and often originates in the oral cavity (68%), followed by the larynx and oropharynx (11% each). The median head and neck squamous cell carcinoma diagnosis age was 31 years, a striking difference from the median age of the general population (60 years old). Diagnosis often occurs at advanced stages (stage IV in 63% of cases), and recurrence is prominent, affecting 49% of patients approximately 22 months posttreatment.^{2,3}

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CASE REPORT

A 37-year-old woman who was diagnosed with FA in childhood presented at our outpatient clinic with pronounced right facial swelling, trismus, and severe pain. Examination revealed a solid oral cavity mass that had developed over 6 months, initially manifesting as minor gingival swelling. Maxillofacial computerized tomography and incisional biopsy confirmed an abscessualized squamous carcinoma of the oral cavity. This carcinoma invaded the mucosa, impacted two teeth, and affected the right jaw (Fig. 1). Following a comprehensive multidisciplinary consultation, radical excision coupled with autologous reconstruction was planned. The procedure encompassed mass excision and the removal of surrounding structures infiltrated by the tumor, including an approximately 8-cm segment of the mandible. By performing an extemporaneous frozen section on the margins and proceeding with ipsilateral modified radical neck dissection, we achieved immediate reconstruction through a single-time, free flap transplantation of the contralateral double-barrel fibula (See Video [online], in which presurgery markings of biaxial fibula flap project are shown, and Fig. 2). The fibula flap design was based on the computed tomography images. A skin paddle was utilized for intraoral lining reconstruction (Fig. 2). Vessel microanastomosis was performed on the facial artery and vein. The postoperative course was uneventful, with the patient remaining hospitalized for 14 days and receiving nasogastric tube feeding. Pathological examination revealed T4N2 staging,

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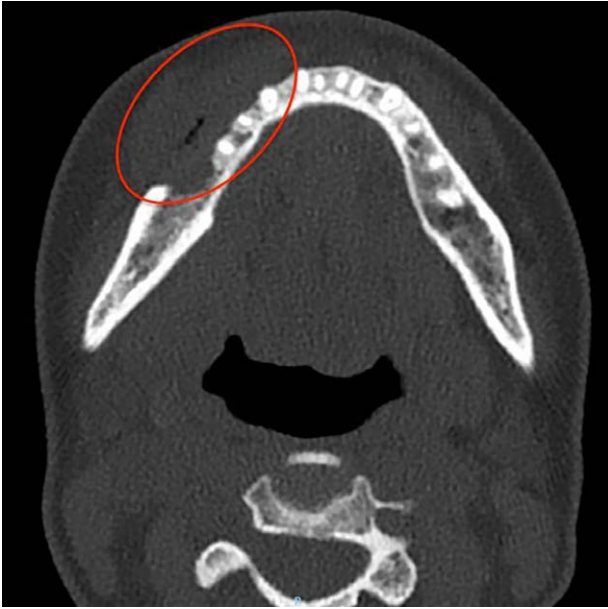


Fig. 1. Presurgery computed tomography. Red circles mark the mass. The tumoral mass eroded the right jawbone.

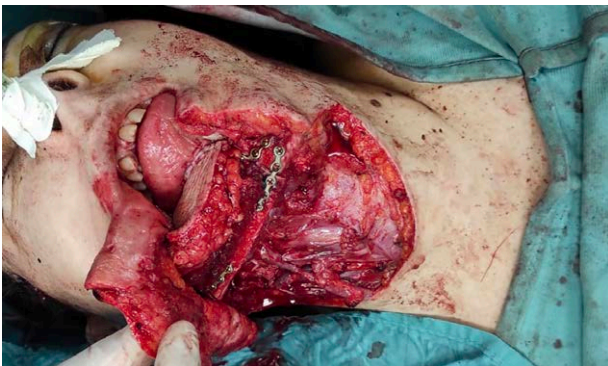


Fig. 2. Insetting phase of the harvested flap to the recipient site.

prompting subsequent radiotherapy (RT) and adjuvant chemotherapy (ChT). Close surveillance was scheduled. A 6-month follow-up revealed no short- or long-term complications, confirming the desired functional and aesthetic outcomes.

DISCUSSION

Head and neck squamous cell carcinomas in patients with FA manifest at a young age, often at an advanced initial stage, and have a marked inclination toward local recurrence. Despite successful surgery with clear margins, swift local recurrence remains a common occurrence in patients with FA, underscoring the delicate balance required to minimize treatment-related morbidity and offer optimal disease management. Patients with FA exhibit compromised DNA crosslink repair capabilities, rendering platinum agents unsuitable owing to heightened toxicity risks. Furthermore, the tolerability of RT was significantly impaired.⁴ Consequently, resection widening

is often necessary, making the reconstruction more complex. Presently, there are no established guidelines for clinical approaches and surgical procedures in these cases. However, the sole sources of information regarding this challenging patient subset are case reports and series.⁵ Although less invasive reconstructive options, such as custom-made implants, are conceivable, the inherent limitations in further dental implantation preclude this approach.⁶ Hence, considering the patient's young age and stable clinical status, we opted for a reconstructive strategy that would yield optimal functional and aesthetic outcomes. The fibula flap, introduced in 1989 by Hidalgo, remains the gold standard for this type of defect reconstruction.⁷ Over time, the free fibula flap technique has undergone several adjustments to accommodate variations in fibula calibers and mandibular remnants. The introduction of the double-barrel fibula flap in 1995 by Horiuchi et al addressed the height discrepancy often encountered in single-barrel mandibular reconstruction.⁸ The biaxial double-barrel fibular flap technique offers robust facial support and a dental implant-friendly alveolar segment. Importantly, it can be executed with minimal additional postoperative morbidity and without extending hospital stay, as seen in single-barrel reconstruction.⁹ The fibula can be harvested purely as an osseous flap or in conjunction with the muscle and skin, rendering it remarkably adaptable for the reconstruction of diverse mandibular and soft-tissue defects. With the peroneal artery predominantly supplying the fibula osteocutaneous free flap and flap drainage facilitated by the two venae comitantes, microanastomosis is straightforward. The average artery caliber was 1.5 mm, whereas the veins measured 3 mm and aligned well with the recipient vessel dimensions in the neck.¹⁰ Notably, the fibula benefits from a dual blood supply, both endosteal and periosteal. Abundant periosteal supply is pivotal for graft survival and facilitates multiple closely spaced osteotomies. This enables the fibula to be easily contoured to reconstruct diverse segmental mandibular defects. Considering these factors, we adopted the double-barrel fibula reconstruction technique, involving a 17-cm fibula harvest and resection of the 1-cm central segment (see **Video 1**). This facilitated 180-degree rotation of the fibula, effectively reconstructing the 8-cm mandibular defect. A skin pad was used for oral cavity lining reconstruction (**Fig. 2**). The 6-month follow-up confirmed both aesthetic satisfaction and mandibular function goals, as shown in **Figures 3** and **4**. No local recurrence was noted, and comprehensive surveillance was scheduled for the subsequent years. Following RT and ChT, dental reconstruction using implants matching the height of the double-barrel fibula and healthy mandible was considered.

CONCLUSIONS

FA, a hereditary DNA repair deficiency, manifests as progressive pancytopenia; bone marrow failure; variable congenital malformations; and heightened susceptibility to hematological and solid tumors, including head and neck squamous cell carcinomas. Consequently, managing these patients poses a challenge given the elevated local recurrence

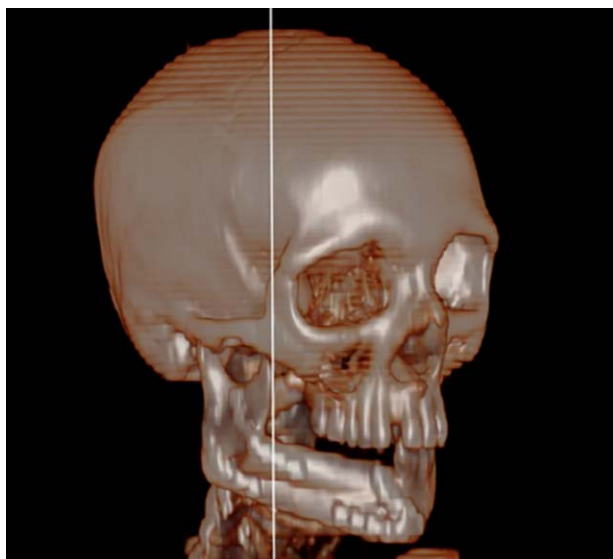


Fig. 3. Computed tomography 3D rendering after 3 months from surgery is possible to see the perfect integration of the osseous flap.



Fig. 4. Three months postsurgery control.

rates and compromised resistance to ChT and RT. This case highlights the complexity of clinical and surgical decisions, emphasizing the necessity of a multidisciplinary approach. The pivotal role of this case in achieving oncological clearance and minimizing aesthetic-functional sequelae is underscored. Reconstruction techniques play a vital role in addressing

these challenges, with the double-barrel osteocutaneous fibula flap emerging as the optimal choice, facilitating future dental implantation without escalating surgical risks.

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DISCLOSURE

The authors have no financial interests to declare regarding the content of this article.

ETHICAL APPROVAL

All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

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