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Sublingual hematoma after autogenous block grafting to the anterior mandible in a patient with Noonan Syndrome

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ABSTRACT

Noonan Syndrome is an autosomal dominant congenital syndrome that is a multisystem disorder, commonly presenting with craniomaxillofacial, cardiac, and hematologic differences. Anterior mandibular surgery presents a surgical challenge because of potential injury to vessels in the floor of mouth. Inadequately-controlled hemorrhage, particularly in the setting of coagulopathy, may quickly escalate to acute airway obstruction. Additionally, modest hypotension maintained under general anesthesia typically reverses upon wakening, potentially resulting in refractory hemorrhage. In this report, we document a case of rapidly evolving floor of mouth elevation secondary to hemorrhage following alveolar symphyseal reconstruction in a patient with Noonan syndrome. Early recognition and operative re-exploration for hemorrhage control, with airway control, administration of high-dose intravenous steroid, and close monitoring in the intensive care unit resulted in rapid resolution of mouth-floor swelling, safe extubation, and a successful surgical outcome.

1. Introduction

Noonan syndrome is an autosomal dominant congenital syndrome with an estimated prevalence of 1 in 1000 to 1 in 2500 live births [1]. It is characterized by abnormal craniofacial growth, congenital heart abnormalities and bleeding disorders [2]. Craniofacial characteristics in Noonan syndrome include widely spaced eyes with down-slanting palpebral fissures, ptosis, and often low-set and posteriorly-rotated ears [2]. Intraorally, these patients may have high-arched palate, speech articulation difficulties, oligodontia, and micrognathia [3,4]. Cardiac anomalies, such as pulmonary valve stenosis, hypertrophic cardiomyopathy, and atrial septal defects are observed in 50–80% of these patients [2]. These patients typically have increased bleeding tendency secondary to thrombocytopenia, platelet dysfunction, and coagulation factor deficiencies. These characteristics increase the risk of significant hemorrhage during surgical procedures [2].

In this report, we present a patient with Noonan Syndrome who presented for extraction of retained primary mandibular central incisors and autogenous block grafting an atrophic anterior mandibular alveolus. Post-operatively the patient developed sublingual hematoma that required urgent return to the operating room for re-exploration and hemorrhage control.

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2. Case report

A 19-year-old 61Kg female patient with a history of Noonan Syndrome presented with an atrophic anterior mandibular alveolus, missing mandibular lateral incisors and retained primary central incisors (Figs. 1 and 2) indicated for extractions and ridge augmentation in anticipation for future dental implant placement. She had undergone a Le Fort I osteotomy and bilateral mandibular sagittal split osteotomies previously without complication. She did not take any medications and had no medication allergies. She had a normal mouth opening. On intraoral examination, her tongue was fully mobile, and the floor of mouth was soft, non-elevated. Her mandibular permanent canines were in place of her missing lateral incisors. She had retained mandibular primary central incisors which were associated with an atrophic alveolus at that site (Fig. 1). Pre-operative panoramic radiograph showed vertical and horizontal bone loss associated with her mandibular primary central incisors (Fig. 2). Her pre-operative lab values were notable for hematocrit 32%, platelet 168K, and INR 1.2. A recent echocardiogram demonstrated mild aortic valve thickening with normal left ventricular size and systolic function.

The patient was taken to the operating room for extraction of the retained mandibular primary central incisors, with immediate reconstruction of the alveolus with an autogenous bone graft harvested from the left mandibular ramus. General anesthesia was induced by facemask (patient preference) using Sevoflurane in nitrous oxide and oxygen. Intravenous access was secured after which a rocuronium bolus was administered to facilitate uneventful fiber-optic nasal-intubation of a size 7mm nasal RAE endotracheal tube. She received 8 mg intravenous dexamethasone shortly after intubation. Sevoflurane in air and oxygen, propofol and dexmedetomidine were used for maintenance of anesthesia. Mean arterial pressures were maintained in the range of 50–60 mmHg throughout the duration of the operation.

Surgery commenced with extraction of the retained primary teeth. A crestal incision was made and buccal and lingual full-thickness mucoperiosteal flaps were raised with no tear in the flaps, revealing a 3mm thin anterior alveolus.

A mucosal incision was then made in the left mandibular vestibule, adjacent to the ramus to gain access to the anterior ramus. A block graft was harvested uneventfully and laid on top of the 3mm thin alveolus. Hemostasis was achieved in the left mandibular ramus donor site with gelfoam and primary closure.

Upon making the initial osteotomy through both the block and the anterior alveolus, the bur appeared to perforate the lingual periosteum. We then encountered pulsatile, bright red bleeding from a vessel in the lingual periosteum. The bleeding was managed with electrocautery, gelfoam, and bimanual pressure for 5 minutes, with complete resolution. The block graft was affixed with a single bicortical screw (Fig. 4). The flap was then closed.

At the conclusion of surgery, the patient was extubated uneventfully and transported to the recovery room in stable condition. Approximately 30 minutes later, it was noted that she had difficulty phonating. Intra-oral examination at that time was notable for an expanding floor of mouth hematoma (Fig. 3A). No acute respiratory distress was observed. The patient was immediately returned to the OR for re-exploration. She was induced without difficulty and intubated nasally. The sutures in the anterior mandible were released and the floor of the mouth was explored. Approximately 10 cc of bright red blood was evacuated, though discreet bleeding vessels could not be readily identified for ligation. Hemostasis was achieved with cautery. The site was packed with hemostatic agents, including absorbable gelatin sponge and fibrin glue and then closed with sutures in a water-tight fashion (Fig. 3B). Blood loss was estimated at 25 mL.

At the conclusion of the procedure, the patient was kept intubated and transferred to the intensive care unit because of concern for swelling to cause progressive upper airway obstruction. Post-operative blood work revealed hematocrit 35%, platelet 188K and INR 1.3 (compared to 32%, 168K, and 1.2 pre-operatively). No blood products were administered.

Overnight, she received 8 mg intravenous dexamethasone scheduled every 8 hours. On post-operative day 1, she had no respiratory distress, passed a spontaneous breathing trial, and had a positive endotracheal tube cuff leak test. Her floor of mouth was less edematous with the palate and oropharynx visible (Fig. 3C and D). She was then extubated uneventfully and received another scheduled dose of dexamethasone immediately after. She was monitored uneventfully for the next several hours and discharged following a final intravenous dose of dexamethasone. Post-operative panoramic imaging showed successful removal of the mandibular

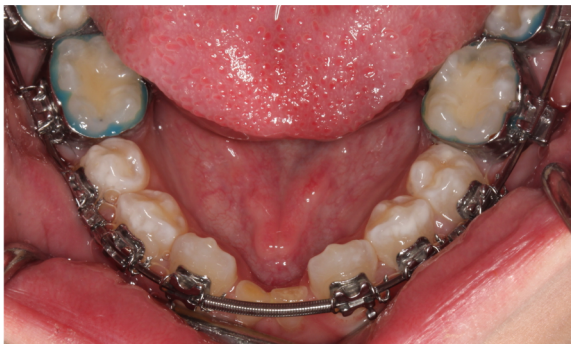

Fig. 1. Pre-operative clinical photo of the floor of mouth. Retained teeth #O&P. The mandibular canines are in position of the absent mandibular lateral incisors.

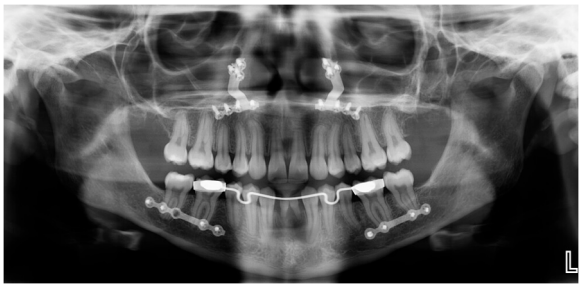

Fig. 2. Pre-operative panoramic radiograph shows hardware from previous LeFort I osteotomy and bilateral sagittal split osteotomy of the mandible for advancement, root resorption on primary teeth #O&P and vertical atrophy of the anterior mandibular alveolus.

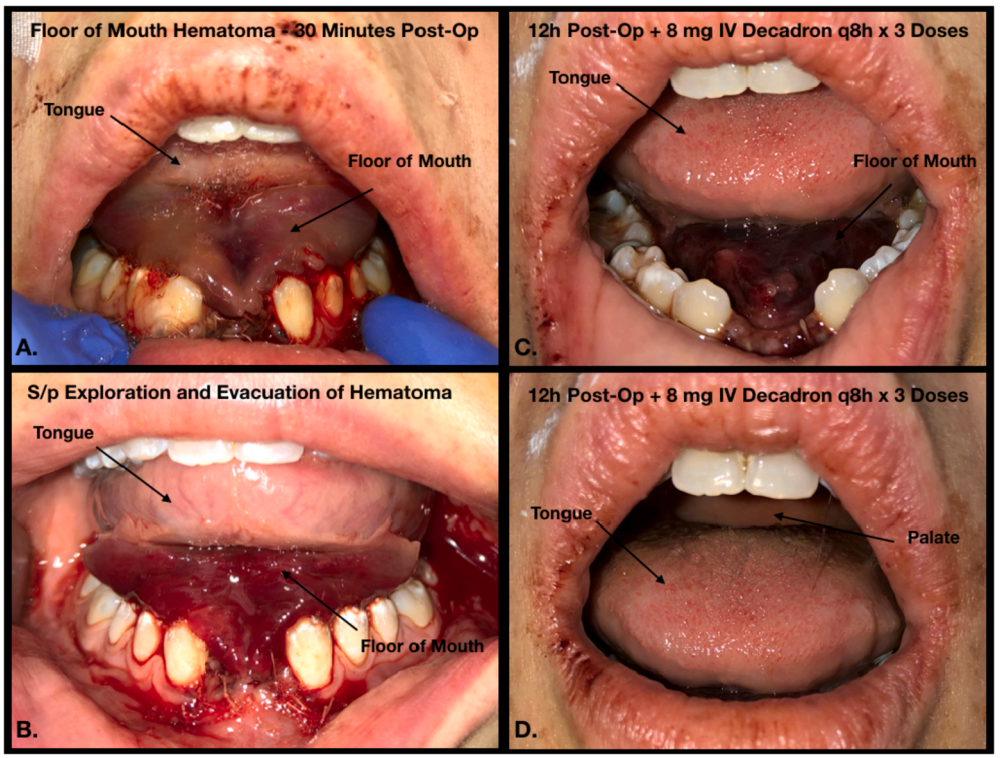

Fig. 3. (A) Floor of mouth elevation, 30 minutes after closure of the anterior mandibular gingival flap (B) Floor of mouth after intraoral exploration and hemorrhage control (C) Floor of mouth and tongue positioning 12h after surgery with 3 doses of 8mg IV dexamethasone (D) Palate can be seen 12h after surgery.

primary central incisors and a block graft secured in the anterior atrophic ridge using a single bicortical screw (Fig. 4).

3. Discussion

The floor of mouth is a highly vascular structure containing muscle, salivary glands, nerves and blood vessels. The important arteries to consider in the floor of mouth attachment to the mandible are the sublingual artery and the perforator branch from the submental artery [5,6]. This patient had a modestly elevated baseline International Normalized Ratio of 1.3. The combination of violation of the anterior mandibular lingual flap, resumption of normal blood pressure upon emergence and baseline coagulopathy may have contributed to the persistent sublingual hemorrhage and subsequent floor of mouth elevation (Fig. 3).

Airway management remains the primary consideration in sublingual hemorrhage. Law, Alam, and Borumandi reported in their 2017 literature review that there were 25 reported cases of floor of mouth hematoma: 84% of these cases were from lingual cortex perforation and 68% required emergency intubation or tracheostomy due to acute airway obstruction [7]. To our knowledge, there are no studies that have defined the incidence of sublingual hematoma following anterior mandibular surgery.

Corticosteroids are potent anti-inflammatory agents, and act by inhibiting leukocyte migration to the area of inflammation,

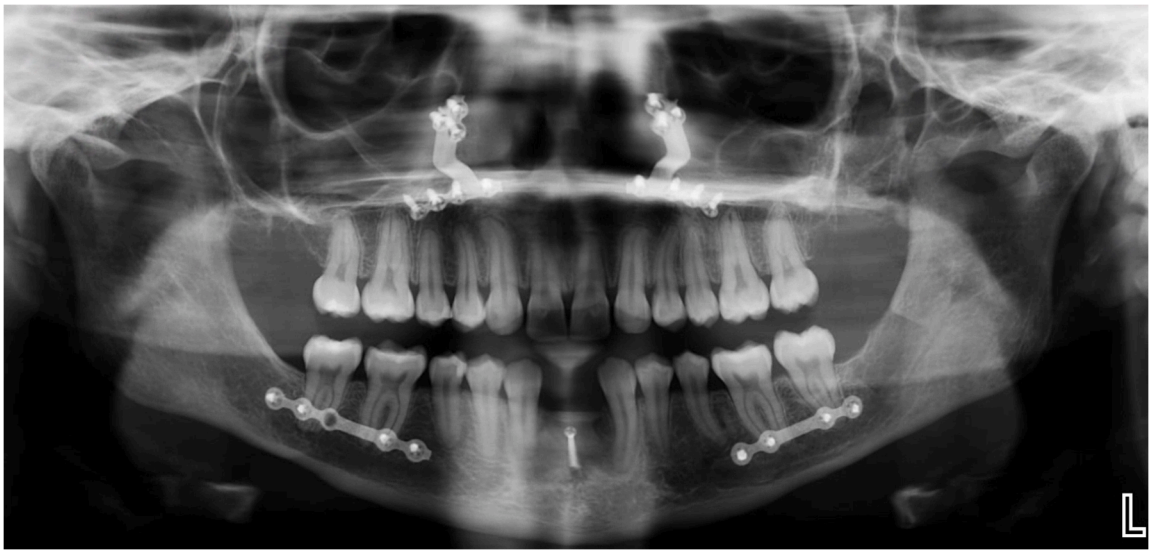

Fig. 4. Post-operative panoramic radiograph shows rectangular radiolucency at the anterior border of the left mandibular ramus, consistent with block graft harvest and mandibular midline block radiopacity with a single bicortical screw, indicating the site of block placement.

decreasing fibroblast and endothelial cell function and suppress the production of cytokines. This results in decrease in vascular dilatation and edema formation [8]. The benefits of pre-operative administration of corticosteroids for reducing postoperative edema has been documented for third molar surgery as well as orthognathic surgery [8,9]. We administered 8mg of intravenous dexamethasone both pre- and post-operatively every eight hours as an adjunctive therapy based on the understanding of its pharmacology and physiology, rather than high level evidence. Given the rarity of post-operative sublingual hemorrhage there is little available evidence to support corticosteroid administration to decrease floor of mouth edema in this context.

Patients with Noonan Syndrome may commonly have dental anomalies, from missing lateral incisors to a combination of missing and unerupted permanent teeth [4,10]. With the emergence of modern dental implant technologies and their increase acceptance by dentists and the public as reasonable treatment modalities for partial edentulism, it comes as no surprise that more people, syndromic or otherwise, will seek this treatment. Elani et al showed that dental implant prevalence in the United States has increased from 0.7% in 1999 to 2000 to 5.7% in 2015 to 2016, with a continued uptrend in the foreseeable future [11].

This case highlights the potential problem that may quickly arise when performing anterior mandibular surgery. The floor of the mouth elevates quickly if hemorrhage is not fully controlled and presents a potentially grave airway risk. Early recognition and intervention is key and airway control is the goal in the immediate perioperative setting.

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Conflicts of interest

None of the authors has a financial or non-financial interest in the products or devices mentioned in this work.

Ethics statement

The guidelines in the Declaration of Helsinki were followed at all times during this work. Institutional review board approval was not required.

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