

Recognizing Osteogenesis Imperfecta as a Risk Factor for Bleeding with Otolaryngology Surgical Procedures: A Case Report of Postoperative Tonsillectomy and Adenoidectomy Bleeding

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Abstract

Osteogenesis Imperfecta (OI), also known as “brittle bone disease”, is a rare autosomal dominant genetic disorder (with 4 subtypes) affecting connective tissue and is characterized by extremely fragile bones that fracture easily often without apparent cause. The genetic mutation has been identified as affecting 1 of 2 genes, *COL1A1* or *COL1A2*, located on chromosomes 17 and 7 respectively. These genes are involved with the production of type I collagen. Collagen is a major protein of bone and connective tissue.

Keywords Osteogenesis imperfecta; Adenoidectomy; Tonsillectomy; Postoperative bleeding

Introduction

In OI, there is a 10-30% incidence of a bleeding diathesis in patients undergoing surgical procedures [1,2]. The coagulation abnormality appears to be associated with the abnormal collagen adversely affecting platelet-endothelial cell interactions and capillary strength. Friable soft tissue with small blood vessels unable to vasoconstrict adequately and a defect in the platelet aggregation response around exposed subendothelium are felt to be a result of the collagen defect in OI. These patients can have increased capillary fragility, decreased platelet retention, decreased levels of factor VIII and deficient collagen-induced platelet aggregation that can be identified through laboratory studies [3]. Unfortunately and even more concerning excessive bleeding can occur despite normal coagulation and platelet function studies.

Desmopressin, an analog of antidiuretic hormone, causes endothelial cells to release von Willebrand's factor, tissue type plasminogen activator and factor VIII:C which promotes platelet adhesiveness to the vascular endothelium, thereby increasing hemostatic activity. This medication is not routinely utilized preoperatively in OI patients, but in anecdotal cases desmopressin has been felt to be useful and even essential in controlling serious postoperative bleeding [2]. This medication can be administered intravenously or topically by nasal spray.

This case report describes a patient with the most common form of OI, type I, which is mildest form [1]. In type I OI most bone fractures occur during childhood through pre-puberty, with a significant decline in bone fractures after puberty. A distinguishing feature of this subtype is the presence of pale “blue sclera”.

Case Report

While on-call the corresponding author was contacted by the emergency department physician regarding a 12-year-old female who

was 11 days post-surgery from tonsillectomy and adenoidectomy and was having significant bleeding from the mouth, presumably from the tonsillectomy sites. Efforts to stop bleeding with ice water gargling were unsuccessful. The patient was also having emesis with bright red and dark blood.

An immediate consultation was performed in the emergency department. Initial examination showed the patient was having continuous bleeding from the oral cavity with some bright red blood at both nostrils. Bleeding started while eating a taco. No history of prior a bleeding problem was identified, including family history. The patient had a left lower extremity cast from a non-traumatic fracture involving her foot.

Although the patient's tonsillectomy and adenoidectomy was performed without difficulty with no intra-operative complications or problems reported by the surgeon, the patient had some nasal bleeding in the Post-Anesthesia Care Unit (PACU) at the Ambulatory Surgery Center (ASC) where she underwent surgery. This was managed successfully with oxymetazoline nasal spray instilled nasally. The patient was discharged without further problems from the ASC. Postoperative medications consisted of hydrocodone/acetaminophen elixir and ibuprofen. No oral steroids were prescribed. Over the following 3 days the patient had intermittent mild bleeding from the mouth that was controlled with ice water gargling. The patient and mother state that no further significant bleeding occurred after 3 days until she presented to the emergency department, but minor bleeding could not be excluded.

The patient's hematocrit in the emergency department was 32.3. Reviewing the patient's clinic chart later showed the patient's preoperative hematocrit was 37.5.

The patient was taken immediately to the operating room where she underwent general endotracheal anesthesia without incident. The adenoidectomy site demonstrated continuous oozing characterized as a welling up of blood over the entire area of the adenoidectomy open wound. The right tonsillectomy surgical site showed a continuous stream of arterial blood from mid-tonsillar fossae region that jetted

across reaching the contralateral lateral pharyngeal wall. Immediate tamponade of right tonsillectomy bed using a radiopaque strung cotton ball packs was performed. Suction monopolar cautery was applied to the adenoidectomy surgical site and 5 separate attempts were required before successful control was achieved. Bleeding at subsequent cauterizations was principally from the peripheral border of the cauterization area. Following control of bleeding from the adenoidectomy surgical site the right mid tonsillar pole small arterial vessel bleeding was controlled with suction monopolar cautery. There was no evidence of infection or unusual inflammation. The Estimated Blood Loss (EBL) for this procedure was 100 mL. EBL for the tonsillectomy and adenoidectomy procedure was reported to be 50 mL after reviewing the operative report later.

The patient tolerated the procedure well and was extubated, followed by transport to the PACU. The patient continued to do well and was transferred to the floor for observation and supportive care with the pediatric hospitalist service.

While convalescing the patient underwent coagulation and platelet function studies which included PT, PTT, platelet count, fibrinogen level, von Willebrand factor, factor VIII activity and platelet function (collagen/epinephrine) all of which were in the normal range except for the platelet count which was abnormally high at 456 (normal range 194-345).

The patient was hemodynamically stable throughout her hospital stay with no blood products given. The patient was discharged the following day. On the day of discharge the patient's hematocrit was 26.7 dropping from a preoperative level of 37.5.

Discussion

The incidence of postoperative tonsillectomy and adenoidectomy bleeding is reported to be approximately 3% and 0.2-0.5% respectively [4,5], with late bleeding from an adenoidectomy site felt to be extremely rare. Osteogenesis imperfecta carries a significant risk of bleeding in patient's undergoing surgical procedures which can be life-threatening or fatal [6]. The risk of bleeding with surgical procedures needs to be appreciated by otolaryngologists as this is a rare disorder

with emphasis on understanding the disorder as it related to bone fractures and not bleeding making it a potential pitfall for otolaryngologists. Furthermore, bleeding can occur despite having normal coagulation and platelet function studies as was found to be true retrospectively with this patient. Preoperative assessment for bleeding disorders while useful if a bleeding diathesis is identified, cannot provide security for the surgeon. Routine pre-operative coagulation screening tests are generally not felt indicated unless a medical or family history of bleeding problems are identified in healthy children undergoing elective tonsillectomy and/or adenoidectomy [7]. Otolaryngologists who encounter a patient with osteogenesis imperfecta who require surgery should seek consultation with hematology oncology knowing that bleeding diatheses are potentially present in 10-30% of patients and there is still an inherent risk of bleeding even with normal studies. Furthermore, knowing that IV Desmopressin may be a useful pharmacologic intervention to help stop bleeding should severe unexpected postoperative bleeding occur, but its use is anecdotal at this time [2]. Having established a patient relationship with hematology oncology is a prudent decision.

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