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An incidental latent adult hemophilia case found after a tonsillectomy

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Abstract

Severe bleeding after a tonsillectomy may cause airway obstruction and be life-threatening. We report post-tonsillectomy bleeding in a 32-year-old patient with hemophilia A, who had not been aware of his disease for more than 30 years. He underwent tonsillectomy for recurrent tonsillitis. He denied episodes of bleeding tendency. The preoperative workup was normal, including platelet count, prothrombin time, and activated partial thromboplastin time. The surgery itself was uneventful, but severe bleeding from the inferior pole of the tonsillar bed developed 7 days after surgery. Emergency hemostasis was performed under general anesthesia in the operating room. The patient then remembered several episodes of bleeding tendency. Coagulation tests revealed a mild lack of coagulation factor VIII to 35%, and a diagnosis of hemophilia A was made. Hemophilia might only be found after surgery and can cause life-threatening complications. However, latent hemophilia detected after a tonsillectomy in a 32-year-old adult is very rare. A careful history of bleeding tendency is important to achieve a diagnosis of coagulopathy, perform a safer surgery, and prevent postoperative complications.

Keywords

Adult, hemophilia A, history taking, postoperative bleeding, tonsillectomy

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Introduction

Tonsillectomy is a popular surgery in medicine particularly in otolaryngology. Indications to perform this surgery are not only recurrent tonsillitis but also obstructive sleep apnea with associated tonsillectomy hypertrophy and other non-otolaryngological conditions such as IgA nephropathy.[1] The surgical technique itself is established, and its risks and complications, such as postoperative bleeding, are well recognized among surgeons.[2,3] Severe bleeding after a tonsillectomy may cause airway obstruction. Therefore, tonsillectomies for patients with bleeding tendencies require careful manipulation, ensuring good intraoperative hemostasis and postoperative care.[4–8] However, such patients who have never been aware of and/or pointed out their bleeding tendency before surgery may exist, and all otolaryngologists should be aware of this possibility.

Case presentation

A 32-year-old male planned to undergo tonsillectomy due to recurrent tonsillitis and snoring. He did not report any bleeding tendency at the first visit, and his preoperative laboratory workup was normal. Blood tests showed a hemoglobin level of 13.8 (normal range, 13.4–17.1 g/dL: all laboratories noted in Table 1), a platelet count of 244×10^9/L ($153–346 \times 10^9$/L), a prothrombin time (PT) of 11.6 s (8.6–14.4 s), and an activated partial thromboplastin time (aPTT) of 32.0 s (21.3–35.5 s).

The surgery itself was uneventful. No significant bleeding was observed, and the total blood loss was 15 mL. The patient started with a soft diet on the day after surgery and gradually returned to a normal diet. Five days after surgery, he noticed a hemosputum. A small blood clot was observed on the right tonsillar bed 6 days after surgery, but no active bleeding was observed.

Eight days after surgery, the patient noticed bleeding after breakfast and visited our hospital. Blood clots and continuous hematic oozing were observed in both tonsillar beds. Hemostasis was obtained under general anesthesia in the

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Table 1. Time course of laboratory data.

Date	Hb	Plt	PT/control	APPT/control	Factor VIII activity
Before surgery	13.8	244	11.6/11.5	32/28.4	NA
POD 2	13.6	188	NA	NA	NA
Postoperative bleeding (POD 8)	12.4	243	12.2/11.4	31.1/28.4	NA
POD 10	9.1	231	12.5/11.5	32.2/28.4	35 (60–150)
Two months after surgery	12.5	237	NA	NA	35 (60–150)
Unit	g/dL	$10^3/\mu L$	s	s	% (normal range)

operating room. Active bleeding was observed on the right tonsillar bed. Only hematic oozing was observed on the inferior pole of the left tonsillar bed. Bleeding was controlled by bipolar electrocautery and ligation. The total intraoperative bleeding was 65 mL. He developed anemia with a hemoglobin level of 9.1 g/dL. Nevertheless, PT and aPTT were still normal (12.5 and 32.2 s, respectively).

After this bleeding event, the patient recalled an episode of recurrent bleeding after a wisdom tooth extraction 10 years previously. After the extraction, slight bleeding had persisted for 4 days. He needed to return to the dentist and underwent hemostat by certain packing. However, bleeding still persisted. He revisited the dentist, and the bleeding finally ceased after gum suturing. Neither other bleeding episodes nor family history of bleeding tendencies was described. According to the Bleeding Assessment Tools score, he had only experienced bleeding episodes from tooth extraction and present surgery and was scored only 6.[9,10]

The bleeding tendency was suspected after the patient reported his bleeding episode after tooth extraction, so coagulation studies were performed. The results revealed a mild lack of coagulation factor VIII activity at 35% (normal values vary between 50% and 150%). Other clotting factors were estimated. Activities of each coagulation factors were within normal range; Factor IX (94%: 70%–130%), Factor X (88%: 70%–130%), Factor XI (82%: 75%–145%), Factor XII (87%: 50%–150%), and von Willebrand factor (88%: 50%–150%). Thrombocytic agglutinability and bleeding time (2.5 min) were also normal. The coagulation factor VIII inhibitor was negative. These factors were assessed again 2 months after surgery, and the same results were obtained. A diagnosis of mild hemophilia A was established.

The patient took a liquid diet for several days, and no more bleeding episodes occurred. He was discharged 4 days after hemostasis control was achieved. We advised him to avoid any kind of trauma and consult with a hematologist before any surgery or procedure with a possible associated bleeding risk. Four months after surgery, the tonsillar beds showed complete reepithelization, and the hemoglobin level returned to normal (13.6 mg/dL).

Discussion

Hemophilia A is an X chromosome-linked bleeding disorder caused by mutations in the coagulation factor VIII genes.[11–13] This factor is involved in the intrinsic pathway of blood coagulation. Affected individuals have severe, moderate, or mild forms of the disease as defined by factor plasma levels of 1% or less, 2%–5%, and 6%–40%, respectively.[12] The prevalence of hemophilia A is 1:5000 male live-births.[13,14] The timing of symptoms differ according to the factor plasma level. In patients with mild forms of this disease, the main cause of bleeding is significant trauma or surgery.[6,13,15–18]

In this case, the patient seemed very healthy and described no disorders other than recurrent tonsillitis in the medical interview. He described no family history of bleeding tendencies, including hemophilia. Nevertheless, he actually had an episode related to bleeding tendency requiring repeated attempts to obtain hemostasis. After this episode, he did not experience other bleeding troubles and had never consulted a medical clinic. As a result, his hemophilia had gone unnoticed for 30 years.

A hemophilia case first found during adulthood is actually rare, and few reports fit this condition. According to Tsuyama et al.,[18] aPTT was normal at the first blood sampling due to the low disease level (1 BU/mL). In such a condition, repeated aPTT tests and measurements of coagulation factors are required. Normal aPTT in the first blood sampling must still be observed for postoperative bleeding. A thorough history might have revealed the bleeding tendency and prevented the present episode of postoperative bleeding.[15,18]

A typical patient with hemophilia has a family history of bleeding tendency and prolonged aPTT, but symptoms differ according to factor plasma levels. Some cases bleed immediately after a tonsillectomy, whereas others do not develop bleeding until 7 days after surgery. Therefore, a patient can bleed after a tonsillectomy more than 7 days after surgery.[6,16] Due to incomplete coagulation, the coat on the tonsillar beds was partially removed and oozing began 5 days after surgery. Patel et al.[8] reported seven children with bleeding disorders involving post-tonsillectomy hemorrhage and in six of the seven children, bleeding episodes were delayed, occurring after 7 days or later. Warad et al.[7] also reported hemorrhagic complications with adenotonsillectomy in children and young adults with bleeding disorders. Bleeding immediately after the surgery was reported, but delayed bleeding (>24 h postoperatively) was more common among children with a bleeding tendency.[4] In these reports, half of the children had never been diagnosed

with a bleeding tendency. Therefore, even patients without bleeding disorders undergoing adenotonsillectomy are at risk of bleeding and require close monitoring with hemostatic support in the postoperative period.[17,19]

Conclusion

Like in this case, hemophilia might be incidentally found after a tonsillectomy, even if the patient did not report any bleeding tendency and the preoperative laboratory workup was normal. Careful history-taking of bleeding tendency is important for safer surgery.

Author contributions

S.Y. and A.T. wrote the abstract, case summary, literature review, and discussion. Y.S. was the surgeon (second hemostat surgery) and performed patient care and data accumulation. A.T. designed the case report, was the surgical instructor, and performed data assessment. N.S. and T.A. performed emergency medical care and helped in drafting the article.

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Ethical approval

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Informed consent

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