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ANAESTHETIC MANAGEMENT OF A CASE OF SEVERE HEMOPHILIA: A CASE REPORT

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KEYWORDS :

INTRODUCTION

Hemophilia A , also known as classical hemophilia is a genetic bleeding disorder caused by insufficient levels of a blood protein called Factor 8. It can present with symptoms ranging from spontaneous bleeding to persistent bleeding after minor trauma or surgery. Here we report a case of an adult with severe Hemophilia A who presented with right submandibular sialadenitis for submandibular gland excision.

CASE PRESENTATION

A 32 year old male presented to the surgery department with complaints of pain and swelling below the right jaw of 4 years duration. A diagnosis of right submandibular sialadenitis was made. The patient was a known case of Hemophilia A since childhood. His Factor 8 assay came to be <1% , with aPTT of 84.8sec. His Factor 8 antibody was found to be negative. He had no previous history of profuse bleeding, hemarthrosis or other comorbidities. The patient was posted for right submandibular gland excision under GA. Hematologist prescribed Factor 8 replacement 3500 IU iv Q24H , and 4000IU iv on the day of surgery 1 hour before the procedure, followed by daily doses upto the 14th postoperative day.

Preoperatively aPTT was 38.7 and platelet count 2.46 lakhs, with remaining investigations within normal limits . In the operation theatre iv line secured with 18G canula and monitors attached. Patient was induce with 2mg/kg Propofol, and 2mcg/kg of Fentanyl.Videolaryngoscopy performed and airway secured with cuffed endotracheal tube No:8 with vecuronium. Minimal airway trauma was assured. Intaoperatively maintained with oxygen, nitrous oxide and isoflurane. Tranexamic acid infusion was given. Vital parameters were stable throughout the procedure and there was minimal intraoperative blood loss.Sugery lasted for 1.5 hours. After surgery patient was extubated and shifted to male surgical icu.Factor 8 continued for 14 days post surgery. Post operative period was uneventful.

DISCUSSION

Hemophilia is a bleeding disorder that slows the blood clotting process. Haemophilia A, also known as classical haemophilia occurs due to low amounts of clotting factor VIII, and haemophilia B, occurs due to low levels of clotting factor IX. Although it is inherited in an X-linked recessive manner, about 1/3 of cases have no previous family history. It affects all races and ethnic groups.Hemophilia A is four times more common than Hemophilia B.In 2011, India reported 14,718 patients with bleeding disorders and 11,586 patients with haemophilia A1.

The severity of bleeding in a patient with hemophilia depends on the level of clotting factor. Normal levels of Factor 8 are 0.5 to 1.5 IU/ml or 50-150%2 (1 IU/ml = 100% of F8 in 1ml of normal plasma) .Table 1 shows the level of clotting factor and the degree of severity of Hemophilia.

Table 1: Level of clotting factor and degree of severity

Severity	Clotting factor level	Bleeding episode
Mild	5- <40% of normal or 5-40 IU/dl	Spontaneous bleeding- rare Severe bleed- only after major surgery or trauma
Moderate	1-5% of normal or 1-5 IU/dl	spontaneous bleeding-occasional; whereas prolonged bleeding- with minor surgery or trauma
Severe	<1% of normal or <1 IU/dl	Spontaneous bleeding into joints or muscles, predominantly in the absence of identifiable hemostatic challenge

Surgery for patients with hemophilia will require additional planning and interaction with the healthcare team than what is required for other patients.Such patients are best managed by a multidisciplinary team in communication with a comprehensive hemophilia treatement centre. Preoperative assessment should include inhibitor screening and inhibitor assay, particularly if the recovery of the replaced factor is significantly less than expected[3]. Adequate quantities of clotting factor concentrates should be available for the surgery itself and to maintain adequate coverage postoperatively for the length of time required for healing and/or rehabilitation.Factor 8 is recommended to be replaced with recombinant or viral inactivated plasma-derived concentrates [6,7].

The dose of factor VIII is calculated by 0.5 × weight (kg) × desired factor level in IU/dl, according to the world federation of hemophilia 2012 (WHF) guidelines[8]. Maximum rate of transfusion should not be more than 3ml/min in adults[9].Plasma FVIII level will raise by approximately 2 IU/dl per unit of transfused F-8/kg of body weight, in the absence of an inhibitor.9 Other pharmacological agents that can be of great value include desmopressin, tranexamic acid and epsilon aminocaproic acid.

Post-operative management of a hemophilia patient should be in a high dependency unit and should be carefully planned. Pain should be managed initially, with intravenous morphine or other narcotic analgesics , followed by an oral opioid such as tramadol, codeine, hydrocodone, and others.When pain is decreasing, paracetamol/acetaminophen may be used.Intramuscular injections should be avoided.A holistic approach should be adopted when managing a patient with hemophilia. Psychological and social support should be provided to the patient and their family.

To conclude we successfully managed a patient with hemophilia with submandibular sialadenitis for submandibular gland excision according to WHF guidelines.

Table 2: Suggested Plasma Factor Peak Level and Duration of Administration[4].

Surgery		No significant resource constraint		significant resource constraint	
		Desired level (IU/dl)	Duration (Days)	Desired level (IU/dl)	Duration (Days)
Major	Pre-op	80-100	-	60-80	-
	Post-op	60-80	1-3	30-40	1-3
		40-60	4-6	20-30	4-6
		30-50	7-14	10-20	7-14
Minor	Pre-op	50-80	-	40-80	-

	Post-op	30-80	1-5	20-50	1-5

Table 3: Definition of adequacy of hemostasis for sugical procedures[5]

Excellent	Intra-operative and postoperative blood loss similar (within 10%) to the non-hemophilic patient. No extra (unplanned) doses of FVIII/FIX/bypassing agents needed AND Blood component transfusions required are similar to non-hemophilic patient
Good	Intra-operative and/or postoperative blood loss slightly increased over expectation for the non-hemophilic patient (between 10 and 25% of expected), but the difference is judged by the involved surgeon/anaesthetist to be clinically insignificant No extra (unplanned) doses of FVIII/FIX/bypassing agents needed AND Blood component transfusions required are similar to non-hemophilic patient
Fair	Intra-operative and/or postoperative blood loss increased over expectation (25–50%) for the non-hemophilic patient and additional treatment is needed Extra (unplanned) dose of FVIII/FIX/bypassing agents factor needed OR Increased blood component (within 2 fold) of the anticipated transfusion requirement
Poor/None	Significant intra-operative and/or postoperative blood loss that is substantially increased over expectation (>50%) for the non-hemophilic patient, requires intervention, and is not explained by a surgical/medical issue other than hemophilia Unexpected hypotension or unexpected transfer to ICU due to bleeding OR Substantially increased blood component (>2 fold) of the anticipated transfusion requirement

REFERENCES

1. World Federation of Hemophilia (WFH) Canada: WFH; 2013. [accessed on May 9, 2013]. Report on the Annual Global Survey 2011.
2. White G, Rosendaal F, Aledort LM, Lusher JM, Rothschild C, Ingerslev J. Definitions in hemophilia. Recommendation of the scientific subcommittee on factor VIII and factor IX of the scientific and standardization committee of the International Society on Thrombosis and Haemostasis. Thromb Haemost 2001; 85: 560
3. Mathews V, Viswabandya A, Baidya S et al. Surgery for hemophilia in developing countries. Semin Thromb Hemost 2005; 31: 538–43.
4. Rickard KA. Guidelines for therapy and optimal dosages of coagulation factors for treatment of bleeding and surgery in haemophilia. Haemophilia 1995;1(S1):8–13.
5. Definitions in hemophilia. Recommendation of the scientific subcommittee on factor VIII and factor IX of the scientific and standardization committee of the International Society on Thrombosis and Haemostasis. JTH 2012 (in press).
6. Evatt BL, Austin H, Leon G, Ruiz-Sáez A, de Bosch N. Haemophilia therapy: assessing the cumulative risk of HIV exposure by cryoprecipitate. Haemophilia 1999; 5: 295–300.
7. Farrugia A. Guide for the Assessment of Clotting Factor Concentrates, 2nd ed. Montreal: World Federation of Hemophilia, 2008.
8. Srivastava A, Brewer AK et al. Guidelines for the management of hemophilia. Haemophilia. 2013;19:e1–47.
9. Hemophilia of Georgia. Protocols for the Treatment of Hemophilia and Von Willebrand Disease, February 2012. (Accessed June 6, 2012) http://www.hog.org/docLib/20090820_HoGProtocol2009.pdf