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Case Report

A Rare Bleeding Disorder and a Precious Surgery: Cochlear Implantation in a Child with Von Willebrand Disease - A Case Report

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Abstract

Von Willebrand disease (vWD) is the most common inherited bleeding disorder with a prevalence of 1 percent [1,2] but only a small proportion of patients come to medical attention because of the mild nature of the bleeding symptoms. The usual clinical presentation is in the form of mucosal bleed, prolonged bleed from minor cuts or trivial trauma or menorrhagia in females [3]. VWD is due to a quantitative or qualitative deficiency in von Willebrand factor (VWF), a complex plasma protein that plays a pivotal role in the control of bleeding by the formation of a platelet plug. Clinical manifestations are highly variable and co-relate with the nature of the deficiency and accordingly, with the management. Here we are reporting one such case of vWD who underwent the major surgery of cochlear implantation.

Keywords: Von Willebrand Disease (vWD); Von Willebrand factor (VWF); Cochlear Implantation

Introduction

Von Willebrand disease (vWD) is the most common inherited bleeding disorder with varied clinical picture. These patients are at risk of hemorrhage when undergoing any major surgery. This case report highlights one such scenario where a patient with vWD underwent a major surgery of cochlear implantation.

Case Report

A 4-year male child presented to our Outpatient department with a history of congenital deafness. Physical examination revealed no significant abnormalities. BERA showed an absence of wave V and an absence of otoacoustic emissions in both ears. HRCT temporal bone and MRI brain studies revealed no structural abnormalities. The patient was diagnosed with bilateral profound sensorineural hearing and planned for cochlear implantation.

During preoperative workup for Cochlear Implantation surgery, he was incidentally diagnosed to have elevated clotting time of more than 30 minutes. Further evaluation of the coagulation profile showed an elevated Activated partial thromboplastin time of 38.7 seconds (normal value being 29.0 - 35.0 seconds).

Elevated APTT was suspected due to either factor 8 deficiency or vWF deficiency. Thus, factor VIII assay and vWF antigen assay were done which revealed quantitative deficiency of vWF along with reduced ristocetin cofactor activity, thus the patient was diagnosed to have type 2 b vWD.

After meticulous planning, the patient was taken up for surgery. Preoperative optimization was done by giving 950 units of vWF and 1 Cryoprecipitate was infused intraoperatively. Post-operatively patient continued to receive Factor infusion, 570 units vWF/day for 2 days, followed by 380 units vWF/day for 5 days. The immediate post-operative period was uneventful until on day five when the patient developed spontaneous nasal bleed. As no local cause of bleed was noted, the patient was started on daily transfusion with cryoprecipitate for another 5 days. The patient was discharged on postoperative day eleven. In total he received, 7,410 units of recombinant vWF and 16 cryoprecipitates.

Discussion

Von Willebrand's disease is the most common inherited bleeding disorder caused by either qualitative or quantitative deficiency of vWF, a clotting factor. Accordingly, the disease manifestations can be classified [4] as follows.

Type 1	Type 2	Type 3
Partial quantitative deficiency of vWF.	Both qualitative and quantitative deficiency of vWF.	Virtually complete deficiency of vWF.
vWF level in blood: 20% - 50% of normal.	Blood levels of vWF may be normal. four subtypes: (depending on the presence and behavior of multimers, molecular chains of VWF) Type 2A, type 2B, type 2M and type 2N	vWF level in blood: < 20% of normal.
Symptoms: mild	Symptoms: mild to moderate	Symptoms: severe, include spontaneous bleeding episodes, often into their joints and muscles.

Table 1: Types of Von Willebrand's disease.

It is diagnosed by quantitative assessment of vWF in a vWF antigen assay and a qualitative assay of function by a glycoprotein (GP) Ib binding assay, a collagen-binding assay, or a ristocetin cofactor activity (RiCof) or ristocetin induced platelet agglutination (RIPA) assays.

Treatment of the condition depends upon the severity of the disease where desmopressin (DDAVP) is the mainstay of treatment [5]. Being a synthetic version of the natural hormone vasopressin, it stimulates the release of vWF from platelets. Other treatment options include replacement therapy with vWF containing concentrates, antifibrinolytic drugs, topical therapy with thrombin or fibrin sealant, and seldom estrogen therapy in some settings in women. DDAVP has multiple routes of administration and the dose vary accordingly. Although effective, a minimum of side effects like tachycardia, vasodilatation, blood pressure variations, and hyponatremia should be anticipated. Various routes and dosages of DDAVP [6] are mentioned in table 2.

	Intravenous dosing	Intranasal dosing	Subcutaneous dosing
Indications	Before invasive procedures Acute episodes of Bleeding	Less serious bleeding not requiring a hospital visit planned minor invasive procedure	Before invasive procedures Acute episodes of bleeding
Dosage	0.3 mcg/kg (maximum 20 mcg)	< 50 kg: 150 mcg > 50 kg: 300 mcg	0.3 mcg/kg (maximum 20 mcg)
	Diluted in 50 ml of normal saline and infused over 20 to 30 minutes.	Spray is administered by metered puffs, each puff containing 150 mcg.	Peak levels are slightly lower and occur later than with intravenousadministration, butother kinetic parameters are similar

Table 2: Routes and dosages for administration of DDAVP.

Other management options include replacement therapy with von Willebrand factor (vWF)-containing concentrates with a goal to maintain vWf activity between 50 - 100% for 3 - 14 days. Commercially available concentrates are a combination of coagulation factor VIII and vWD. An intravenous infusion over less than 20 minutes at 8 to 24-hour intervals with a dose of 20 to 50 IU/kg is required to keep vWF levels above 50% or to control clinical bleeding.

In case of severe active haemorrhage, cryoprecipitate at a dose of 1 unit per 10 kg body weight can be given, however its use should be limited to only emergency settings. As an adjuvant, antifibrinolytic agent like Epsilon aminocaproic acid (EACA) and tranexamic acid can be administered orally or intravenously. They act by preventing dissolution of the haemostatic plug. In female patients, oral contraceptive with estrogen are also implicated as they act by inducing synthesis of vWF [7].

Learning Points/Conclusion

Von Willebrand's disease is the most common inherited bleeding disorder and it can have varied clinical manifestations ranging from mild to severe bleeding symptoms. Few patients can remain asymptomatic for long. Thus, a routine assessment of bleeding parameters should be done before any major surgery to avoid any perioperative complications. Our patient was meticulously diagnosed and managed by the operating surgeons and the precious surgery was successfully completed without any complications.

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Conflict of Interest

None to declare.

Informed Consent

Not applicable.

Author Contributions

R.K is a major contributor in writing the manuscript, table design and data interpretation. AK and SP participated in writing and editing. AA is the operating surgeon and a major contributor in the design and writing of the manuscript. All authors read and approved the final manuscript.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

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