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Case Report

Recurrent Otorrhagia in a Young Girl: A Diagnostic Dilemma

Iqbal KMM[1], Eldhose C[2], Ali FM[3] and Jalal MJA[4]*

[1]Department of Internal Medicine and Rheumatology, VPS Lakeshore Hospital, Kochi, Kerala, India

[2]Department of Internal Medicine and Rheumatology, VPS Lakeshore Hospital, Kochi, Kerala, India

[3]Department of Cardiology, Government Medical College, Trivandrum, Kerala, India

[4]Department of Internal Medicine and Rheumatology, VPS Lakeshore Hospital, Kochi, Kerala, India

***Corresponding author:** Muhammed Jasim Abdul Jalal, Department of Internal Medicine and Rheumatology, VPS Lakeshore Hospital, Kochi, Kerala, India

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Abstract

A 15 year old female presented with recurrent episodes of otorrhagia lasting for few minutes. She gives history of 1 episode of epistaxis in the past, which resolved spontaneously. She denied any joint pains and easy bruising. There was no medical history of tinnitus, vertigo, otalgia, or otorrhea, or any history of trauma or surgery of the head and neck. She denied history of any allergies. There was no family history of any hemorrhagic disease. She was hemodynamically stable. Ear, nose and throat examination was normal. The patient was evaluated by multiple otorhinolaryngologists, but could not find a cause for recurrent ear bleeding and hence it was a diagnostic challenge. All the laboratory parameters including hemogram, liver function and renal functions were normal. Coagulation parameters - PT, aPTT, Fibrinogen and Platelet count were normal. Since all investigations were within normal limits, a clinical suspicion of von willebrand disease was kept in mind.

Keywords: Otorrhagia; epistaxis; Desmopressin

Introduction

Von Willebrand Disease [VWD] is the most common congenital bleeding disorder. A prolonged bleeding time with a normal platelet count is the most important laboratory abnormality. In 1926, von Willebrand described a novel bleeding disorder. Only in the 1950s, it was demonstrated that the prolonged bleeding time is associated with reduced Factor VIII. In 1970s, it was clarified that the deficiency of a new factor, called Von Willebrand Factor [VWF] and different from Factor VIII, was actually responsible for the disease. VWF is essential for platelet-sub endothelium adhesion and platelet-to-platelet interactions as well as platelet aggregation in vessels. VWF is the specific carrier of Factor VIII in plasma, thus protecting it from proteolytic degradation, prolonging its half-life in circulation and efficiently localizing it at the site of vascular injury.

Type 1 VWD is the most common form, accounting for up to 80% of reported cases, and is generally transmitted as an autosomal dominant disorder. Mucocutaneous bleeding is the most common symptom in these patients, and typically presents as epistaxis, easy bruising, menorrhagia, gingival bleeding and posttraumatic or postsurgical bleeding [1]. Recurrent otorrhagia have been reported secondary to trauma such as self-instrumentation, catheterization, falls or straddle injuries [2,3]. In this report, we describe a patient with unrecognized VWD, in whom the presenting symptom was recurrent otorrhagia.

Case Presentation

A 15 year old female child presented with recurrent episodes of otorrhagia lasting for few minutes. She gives history of 1 episode of epistaxis in the past, which resolved spontaneously. She denied any joint pains and easy bruising. There was no medical history of tinnitus, vertigo, otalgia, or otorrhea, or any history of trauma or surgery of the head and neck. She denied history of any allergies. There was no family history of any hemorrhagic disease. The patient was evaluated extensively, but could not find a cause for recurrent ear bleeding and hence it was a diagnostic challenge.

A detailed clinical examination was done. The child was hemodynamically stable with a heart rate of 84/minute, respiratory rate of 24/minute and blood pressure of 114/78 mmHg. Ear pinna was normal. There was no evidence of ear discharge, wax or foreign body. Tympanic membrane was intact without any perforation. There was no tragal/mastoid tenderness. Audiogram was normal. Nasal tract was clear without any congestion. Nasal septum did not show any deviation/perforation. Paranasal sinuses were normal and non-tender. Diagnostic nasal endoscopy was normal except for adenoid hypertrophy. CT PNS was unremarkable. Lips, Oral cavity, tongue, teeth and gums did not show any evidence of active/passive bleeding. All other systemic examinations were within normal limits.

All the laboratory parameters including hemogram, liver function and renal functions were normal. Coagulation parameters - PT, aPTT, Fibrinogen and Platelet count were normal. Since all investigations were within normal limits, a clinical suspicion of VWD was kept in mind. VWF was 25 (normal values being in between 49 and 187 U/dl). Factor VIII was 28. 4 (normal values being in between and 187 U/dl). Both VWF and Factor VIII values were low in our patient. Hence, she was diagnosed as type 1 VWD with factor VIII deficiency. She was started on a fixed daily dose of 200mcg desmopress in metered-dose intranasal spray.

Discussion

VWF is a large multimeric glycoprotein present in plasma. It is synthesized in Weibel-Palade bodies in endothelium, α-granules of platelets (megakaryocytes) and sub-endothelial connective tissue. VWD is a diagnostic entity recognized for at least 80 years, and considered the most common inherited disorder. Mutations at

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the VWF locus on chromosome 12 leads to VWD [4,5]. Although generally considered to be inherited as an autosomal dominant trait, VWD inheritance may be more complex, including compound heterozygosity, which may contribute to phenotypic variability seen in the disease [6]. Further, extragenic factors including ABO blood groups [7], adrenergic states [8], hormones [9] and inflammatory states [10] may cause fluctuation in VWF levels and mask a VWD diagnosis.

According to International Society on Thrombosis and Haemostasis guidelines 2006,

VWD is classified into 3 types:

Type 1- Mild/moderate deficiency of qualitatively normal VWF (Autosomal dominant)

Type 2- Qualitative mutants

1) Type 2A: reduced platelet-dependent function with abnormal multimers

2) Type 2B: increased affinity for platelet binding

3) Type 2M: reduced platelet-dependent function with normal multimers

4) Type 2N: reduced FVIII binding

(2A, 2B and 2M are autosomal dominant, whereas 2N is autosomal recessive)

Type 3- Severe deficiency of VWF (Autosomal recessive)

Diagnostic symptoms of VWD are:

- Easy bruising
- Prolonged bleeding from lacerations
- Epistaxis
- Bleeding from gums
- Menorrhagia
- Post-dental procedural bleeding
- Post-surgical bleeding
- Excessive post-partum bleeding
- Muscle hematomas (type 3 VWD)
- Hemarthroses (type 3 VWD)

The goal of the treatment of VWD is the correction of the low FVIII: Cand of the low VWF. Desmopressin and transfusion therapy with blood products represent the two treatments of choice in VWD [11]. Other forms of treatment can be considered as adjunctive or as an alternative to these two modalities. Desmopressin is a synthetic analog of vasopressin that elevates FVIII and VWF plasma concentrations in most patients with mild hemophilia A and VWD. It is relatively inexpensive, carries no risk of transmitting blood-borne viruses, and can be used for home treatment. Desmopressin is usually administered at a dose of 0.3 μg/kg body weight by a subcutaneous or intravenous route or at a fixed dose of 150 to 300 μg by a metered-dose intranasal spray. Patients with type 3 VWD are usually unresponsive to desmopressin, although some patients do experience an increase of FVIII: C to effective hemostaticlevels, even without a change in the Bleeding Time (BT). Therapeutic infusions can be repeated every 8 to24 hours, depending on the type and severity of the bleeding episode.

Conclusion

Recurrent otorrhagia is very rare in the general population, and we did not find in the literature a description of cases of otorrhagia as a presenting symptom of VWD. When recurrent ear bleeding occurs, it is mild and in most cases the symptoms resolve spontaneously without the need of invasive treatment. We decided to publish this unusual case report of a patient presenting with recurrent otorrhagia, since we want to draw attention to the presence of hematologic disorders in cases of unexplained recurrent otorrhagia. We stress the importance of including the evaluation for existing coagulopathies, such as VWD, in such cases.

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