SHORT REPORT

von Willebrand Disease Associated with Superficial Temporal Artery Pseudoaneurysm

B.J. Ricciardo,¹ B.P. Mwipatayi,⁎ M. Abbas,¹ K. Sieunarine¹ and J.W. Eikelboom²

Departments of ¹Vascular Surgery, Royal Perth Hospital, and ²Haematology, School of Medicine and Pharmacology, University of Western Australia, Perth, WA, Australia

We report on two patients with von Willebrand disease (vWD) that presented with superficial temporal artery pseudoaneurysms following minor blunt trauma. We discuss the possible pathophysiological link between vWD and blood vessel abnormalities. The cases highlight the importance of considering the diagnosis of vWD in patients presenting with pseudoaneurysm.

Keywords: Pseudoaneurysm; von Willebrand disease; Collagen; Temporal artery.

von Willebrand disease is a common bleeding disorder characterised by deficiency (types I and III) or dysfunction (type II) of von Willebrand factor (vWF). Type I vWD, constituting 70% of cases,¹ is often difficult to diagnose as affected individuals have mild non-specific bleeding symptoms.² It is important for surgeons to recognise low vWF levels as a marker of increased bleeding risk that can be readily treated with simple and effective therapies.

An association has previously been reported between vWD and pathological conditions that involve the vessel wall, including arterial aneurysms,³ arteriovenous malformations,⁴ gastrointestinal angiodysplasia⁵ and telangiectasia.⁶ We report two cases of superficial temporal artery pseudoaneurysms following relatively minor trauma in patients with vWD who were successfully treated with surgery. We also discuss the pathophysiological link between vWD and pseudoaneurysm.

Case Reports

Case 1

A 16-year-old boy, with known mild type 1 vWD diagnosed on the basis of bleeding and positive family history, presented with a 2 month history of mild discomfort and swelling in the right temporal region after being struck on the head with a softball. The swelling was not increasing in size and was primarily of cosmetic concern. Examination revealed two discrete, pulsatile masses over the lateral forehead. Ultrasound demonstrated two pseudoaneurysms involving the right superficial temporal artery measuring 18 and 10 mm in diameter.

Following administration of pre-operative desmopressin (DDAVP, 0.3 mcg/kg), the patient was taken to surgery and the two pseudoaneurysms were excised. One pseudoaneurysm subsequently recurred and was successfully re-excised under DDAVP cover with no further recurrences. There were no bleeding complications.

Case 2

A 14-year-old girl presented with a 4 weeks history of swelling in the left temporal region following a fall
while skating, she had no personal or family history of bleeding. Examination revealed a discrete, compressible, pulsatile mass over the left lateral forehead. Ultrasound demonstrated a pseudoaneurysm involving the anterior branch of the left superficial temporal artery measuring 8 mm in diameter. Prompted by our experience in case 1, we performed laboratory testing that demonstrated borderline factor VIII complex levels, consistent with a diagnosis of mild type 1 vWD (Table 1). The patient was taken to surgery where the pseudoaneurysm was successfully excised. There were no bleeding complications.

Discussion

A review of the literature reveals that 75% of cases of superficial temporal artery aneurysm/pseudoaneurysm are associated with blunt trauma. The superficial temporal artery is particularly susceptible to pseudoaneurysm formation following trauma because of its proximity to the frontal bone and minimal overlying muscular cushioning. Surgical ligation of afferent and efferent vessels and excision successfully treats the pseudoaneurysm in most cases.

Reduced or dysfunctional vWF provides conditions that favour pseudoaneurysm formation by prolonging bleeding following damage to a vessel wall. To our knowledge, an association between vWD and pseudoaneurysm of the superficial temporal artery has not been previously reported. However, our observations are consistent with previous reports of an association between vWD and pseudoaneurysm of vessels at other sites (supra-orbital, hepatic, femoral, and superior gluteal arteries), and suggest a common pathophysiologic link between vWD and defects of the blood vessel wall.

von Willebrand disease is the most frequent inherited bleeding disorder, with a reported prevalence of 1–2%.8 The risk of bleeding of patients with low vWF can be readily reduced with a pre-operative infusion of intravenous DDAVP that rapidly increases plasma levels of vWF and has been used clinically for more than 25 years to prevent or treat bleeding in patients with vWD or mild haemophilia.

In the event of vessel trauma, endothelial cells secrete vWF and also deposit vWF on the abluminal surface of blood vessels in close association with the internal elastic lamina. In vitro studies suggest that vWF interacts directly with collagen and there may be a genetic relationship between the production of factor VIII and type III collagen. Collagen is a key component of the vascular basement membrane and reduced or absent levels of type III collagen may be implicated in the pathogenesis of aneurysm formation. Collagen studies on the histological specimens would have allowed further evaluation, but were not available. More data on the possible link between type III collagen deficiency and pseudoaneurysm formation in patients with vWD is needed.

The diagnosis of vWD in patients undergoing surgery is important as there are now simple, readily available, and effective treatments that can reduce the risk of bleeding (e.g. DDAVP, coagulation factor concentrates).

We suggest that surgeons should consider the diagnosis of vWD and measure vWF levels in all patients who present with pseudoaneurysm, even in the absence of a personal or family history of bleeding, in order to identify patients who may benefit from the administration of DDAVP to minimise the risk of bleeding complications. Meanwhile, further work is required to better understand the link between vWD and the pathogenesis of this disorder.

References


Table 1. Results of laboratory testing for vWD

<table>
<thead>
<tr>
<th>Laboratory test</th>
<th>Case 1</th>
<th>Case 2</th>
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<tr>
<td>INR (normal range [NR] 0.9–1.3)</td>
<td>1.2</td>
<td>1.2</td>
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<tr>
<td>APTT (29.5–40.5 s)</td>
<td>41.9</td>
<td>46.5</td>
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<tr>
<td>APTT 50:50 NP</td>
<td>37.0</td>
<td>37.7</td>
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<tr>
<td>Fibrinogen (2.0–4.0 g/L)</td>
<td>4.0</td>
<td>2.8</td>
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<tr>
<td>Factor VIII (50–200%)</td>
<td>50</td>
<td>55</td>
</tr>
<tr>
<td>Ristocetin CoF (50–200%)</td>
<td>50</td>
<td>62</td>
</tr>
<tr>
<td>VWF-antigen (50–170%)</td>
<td>75</td>
<td>80</td>
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Borderline factor VIII complex levels and ristocetin cofactor (CoF) in both patients is consistent with a diagnosis of mild type 1 vWD (von Willebrand disease). In conjunction with factor VIII coagulant activity and factor VIII antigen, ristocetin cofactor is useful for diagnosis of vWD.


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