

The Phenomenon of External Pressure Rumpel Leede Sign: A Review

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Abstract

Rumpel leede sign (RLS) is a clinical presentation observed at the extremities due to pressure applied externally. The appearance ranges from scattered pin-point rashes to an entire arm covered with petechial hemorrhage depending upon the severity. This condition occurs mainly due to fragility of the capillaries which in turn is caused by underlying conditions like hypertension, diabetes mellitus, infections caused by certain organisms, drug-induced RLS etc. When a patient with such condition experiences an external pressure i.e. application of a tourniquet, sphygmomanometer cuff inflation, the region distal to the applied pressure displays the sign. It's most commonly seen in elderly patients and sometimes appears on the thighs of babies when the baby carrier straps are too tight. This phenomenon is self-limiting and thus does not require additional treatment. It resolves within a period of hours to days, sometimes weeks depending on how severe is the presentation. The RLS is also used as one of the diagnostic criteria to identify disease conditions like dengue.

Keywords: Pressure; Capillaries; Pin-point; Severity

INTRODUCTION AND BACKGROUND

Rumpel Leede (R-L) sign is a rare occurrence where petechial rashes are observed due to an external pressure. This phenomenon materializes due to the rupture of dermal capillaries in the peripheral region distal to the pressure. Application of a tourniquet, inflation of the sphygmomanometer cuff are certain ways to trigger the event (1). This sign can be used as a diagnostic method to assess the capillary fragility (2). It is associated with hypertensive vascular disease, thrombocytopenia and diabetic microvascular fragility (3). This clinical sign is mostly predisposed in diabetic patients with increased capillary fragility and high prevalence of hypertension (4,5).

The Rumpel Leede phenomenon was first portrayed by Dr. Theodor Rumpel in 1909 followed by Dr. Carl Stockbridge Leede in 1911 while managing patients with scarlet fever. Pinpoint non blanching spots were observed distal to the application of a tourniquet (2). The R-L sign is presented as mostly pinpoint rashes which can vary from patients to patients based on the severity of the underlying disease. There are no complications succeeding this phenomenon as it resolves spontaneously when the triggering factor is eliminated.

Epidemiology

A study of capillary fragility in diabetes had ascertained that 68% of 72 diabetic patients in comparison with 35% age and sex matched non-diabetic controls showed a positive Rumpel- Leede sign (4,5,6). An exceptionally higher incidence of positive fragility tests is seen in older patients with Diabetes mellitus concurred with atherosclerotic vascular diseases and hypertension (7). The event is positively correlated with diabetic microvascular complications like neuropathy, nephropathy, retinopathy and duration of diabetes. R-L sign can appear with other causes of increased capillary fragility such as Ehlers-

Danlos syndrome, thrombocytopenia and therefore it is not particular to diabetes (4,5,6).

Clinical presentation

The most eminent clinical sign is a well differentiated petechial rash distal to the sphygmomanometer cuff (8-13). Multiple cutaneous petechiae is a positive sign for R-L phenomenon (14). Non-palpable purpura with multiple petechiae of which some had progressed to ecchymosis was also one of the signs of R-L sign (12-17). Striae with petechial rash was also reported in one of the case reports (1). Small non-palpable non-blanching erythematous and violaceous macules below the area of the application of the tourniquet was observed in one of the cases (10). The signs are usually benign and gradually resolves completely over 3-14 days without any serious consequences (3,10,11,15,18).

Risk factors

R-L event is believed to occur most commonly in patients with underlying vascular conditions which could predispose them to vasculopathy such as hypertension, diabetes mellitus and Ehlers-Danlos syndrome (1,13,18). This phenomenon is elicited in diseases which are correlated with increased capillary fragility such as disseminated intravascular coagulopathy, meningococemia, rocky mountain spotted fever (RMSF), fat embolism, thrombotic thrombocytopenic purpura and idiopathic thrombocytopenic purpura (9). Non-invasive blood pressure monitoring and old age are also crucial determinants for Rumpel- leede sign (3,15). R-L phenomenon has also been associated with conditions such as dengue fever, Epstein-Barr virus infection, scarlet fever, Zika virus, after restraint for electroconvulsive therapy and following radial artery cannulation for per cutaneous coronary intervention. In children bleeding diatheses such as idiopathic thrombocytopenic purpura (ITP) and Henoch Schönlein purpura are causative factors for R-L sign (19).

Most recently, tourniquet like forces associated with baby carriers has been reported as a cause for R-L phenomena.

Mechanism

R-L sign is considered relevant to decreased number of platelets and vasculopathy that can dispose patients to vascular fragility (4, 20). Diabetes Mellitus, hypertension, Ehlers-Danlos are the major contributory factors for vasculopathy (20). This phenomenon is also associated with thrombocytopenia and IV drug use (2-5).

Diabetes

Microangiopathy is the inducing element for capillary fragility in patients with long standing diabetes mellitus (2,4,11). The exact mechanism of developing diabetic microangiopathy is incompletely understood but probably influence of genetics has a role (21). Biochemical, ultrastructural, hemostatic and functional changes such as increased protein glycation, abnormal blood viscosity increased organ blood flow, abnormal endothelial function, abnormal platelet function, capillary basement membrane thickening increased vascular permeability, possibly increased free radical activity, increased flux through the polyol pathway and hemostatic abnormalities may interact. The hallmark feature of diabetic microangiopathy is the abnormal thickening of the capillary basement membrane (21,22).

Generalized microvascular vasodilation due to multiple factors such as chronic plasma volume expansion, altered levels of vasoactive substances, tissue hypoxia and altered vasomotor responsiveness which eventually can ensue in vasodilation. Consequent elevation in capillary pressures and flows along with this vasodilation, may be the initiating mechanism leading to both renal and extra renal diabetic microangiopathy (23). Hyperglycemia appears to be a central feature in diabetic microangiopathy which culminates in tissue ischemia (22,24). Duration of diabetes along with the presence of microvascular complications of diabetes including nephropathy, retinopathy and neuropathy has a positive correlation with R-L phenomenon (5).

Hypertension

Increased venous pressure in hypertensive states especially during blood pressure cuff inflation can result in the development of R-L sign (2,11,13). Significant increase in systolic venous pressure can result in rupture of dermal capillaries (1,12). There is a case report which describes Rumpel-Leede phenomenon after hemostasis with a radial artery pressure band after coronary angiography (16). Blood flow restriction training associated with tourniquet like forces of baby carriers has also been reported as a rare cause for R-L sign. This phenomenon was reported in patients with impaired collagen integrity leading to capillary fragility and after prolonged venous occlusion by a tourniquet.

Pediatrics

Purpura in healthy infants (baby carrier purpura) due to tourniquet like forces was recently reported (19).

Drug- Induced

R-L sign was observed in a patient treated with aspirin and prasugrel mainly due to the anti-platelet activity of the drugs (13). There are three reports on amlodipine induced thrombocytopenia which would be a risk factor for R-L sign (15).

Diagnosis

An extreme extent of petechial rash (also petechial hemorrhage) can be considered as the R-L sign and does not need further diagnosis. However, the R-L sign itself can be a diagnostic marker for certain disease conditions (2). The Tourniquet test, also known as the R-L capillary-fragility test is an approach to assess the capillary fragility. WHO has marked the Tourniquet test for the diagnosis of dengue.

Management

The R-L sign is not an alarming condition and is rarely conjoined with pain and discomfort. No definite treatment is available apart from treating the underlying complications like thrombocytopenia or any vascular diseases.

CONCLUSION

Rumpel-Leede phenomenon can be dangerous sometimes, hence it should not be ignored. More awareness should be created among health-care professionals.

Conflict of interest

The authors declare none.

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