CASE REPORT

Rumpel-Leede phenomenon in a patient with adult-onset Still's disease

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SUMMARY

Rumpel-Leede phenomenon (RLP), also known as acute capillary rupture syndrome (ACRS), is a rare occurrence where distal dermal capillaries rupture in response to a proximal compressive force, such as a blood pressure cuff or tourniquet. This phenomenon has been reported to occur in states of vascular fragility such as long-term steroid use, hypertension or diabetes mellitus. Here, we provide a report of RLP occurring secondary to tourniquet application in a 26-year-old woman with adult-onset Still's disease (AOSD) and a recent drug rash. In this case, the cause of the phenomenon is most likely multifactorial. Likely contributing factors include long-term steroid use for the treatment of AOSD, and increased vascular permeability secondary to the drug rash. Patients and clinicians should be aware that the treatment of AOSD may induce a state of capillary fragility and they should work together to minimise the risk of complications.

BACKGROUND

This is the first report of Rumpel-Leede phenomenon (RLP) occurring in a patient with adult-onset Still's disease (AOSD). The case is of educational interest due to the rarity of the phenomenon and its occurrence in conjunction with a rare rheumatological disease. Furthermore, this case lends credence to the suggestion that special consideration should be given to this complication in patients with autoimmune diseases which require long-term steroid use.¹ Clinicians and patients should be aware that RLP can occur in patients with AOSD. The patient discussed in this report has reviewed the content and provided written consent for publication.

CASE PRESENTATION

Our patient was a 26-year-old woman with a 5-year history of AOSD. She presented to the emergency department after developing a diffuse, intensely pruritic rash on her last day of a 5-day course of Bactrim which she had been prescribed for a urinary tract infection (UTI) (figure 1). The diagnosis of drug rash was made. The patient was admitted for observation and supportive care. The patient was begun on high-dose methylprednisolone and antihistamines. The patient reported that she had been on prednisone 4 mg, oral, daily for the past 2 years for control of her AOSD.

During the course of her admission, a tourniquet was applied to the patient's right upper extremity in preparation for a routine intravenous placement. While identifying a vein for access, the nurse noted that a new rash was beginning to appear distal to the tourniquet. The nurse promptly removed the tourniquet and alerted the medical team. The tourniquet was in place for a total duration of <1 min before the procedure was aborted. A unilateral petechial rash was observed on the right upper extremity distal to the site of the tourniquet application, extending into the hand with a sharply demarcated proximal border (figure 2). The rash itself, while seeming to overlie the fading urticarial macules, was clearly distinct from her original rash. On examination, pulses were intact, capillary refill was normal, and sensation and strength were preserved.

INVESTIGATIONS

Pertinent labs drawn that day were as follows:

- ▶ White blood cell (WBC) count 7.23 (4.80–10.80)
- ▶ Haemoglobin 10.03 (12.0–16)
- ▶ Haematocrit 31.0 (37.0–47.0)
- ▶ Platelets 178 K/ μ L (130–400 K/ μ L)
- ▶ Alanine aminotransferase (ALT) 32 (10–50)
- ▶ Aspartate aminotransferase (AST) 51 (15–50)
- ▶ C-reactive protein (CRP) 158.3
- ▶ Erythrocyte sedimentation rate (ESR) 22

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Figure 1 The patient's drug rash prior to application of a tourniquet and the development of Rumpel-Leede phenomenon.

Figure 2 Rumpel-Leede phenomenon.

- ▶ Fibrinogen 254 (180–500)
- ▶ Ferritin 698
- ▶ Lactate dehydrogenase (LDH) 1372 (300–600)
- ▶ Triglycerides 134 (10–140)
- ▶ Prothrombin time (PT) 14.6 (12.3–15.0)
- ▶ Partial thromboplastin time (PTT) 29.3 (24.3–37.2)
- ▶ International normalized ratio (INR) 1.14.

DIFFERENTIAL DIAGNOSIS

The labs were unchanged from her AOSD baseline and the platelet and coagulation studies were within normal limits.^{2 3} The normal laboratory values and physical examination findings combined with the setting in which the skin findings occurred led to the diagnosis of RLP. Thrombocytopenia, vasculitis, allergic reaction to the tourniquet material and worsening drug rash were all considered as potential aetiologies; however, they were rejected in favour of RLP as they did not fit the clinical picture.

TREATMENT

The patient's main concern with the RLP was long-term cosmesis. The patient was reassured that the rash would resolve spontaneously over the next 1–2 weeks. No specific treatment was indicated or initiated for the RLP. Supportive treatment was continued for her admitting diagnosis of drug rash. A follow-up appointment with the patient's rheumatologist was made on the patient's behalf.

OUTCOME AND FOLLOW-UP

The patient followed up with her rheumatologist 1 week later and by that time the petechial/purpuric rash from RLP had completely resolved (figure 3). The patient was left with no long-term complications.

DISCUSSION

RLP has classically been described in the settings of thrombocytopenia,⁴ hypertension⁵ and diabetes mellitus.⁶ There have also been reports of the phenomenon occurring in the settings of chronic steroid use⁷ and Sjogren's disease.¹ The common themes, regardless of aetiology, are capillary fragility and increased intracapillary pressure. In this patient, the capillary fragility was likely multifactorial. Patients with AOSD frequently require long-term steroid therapy. This patient had been on 4 mg of prednisone daily for 2 years. Chronic steroid use alters the synthesis of collagen and therefore leads to a state of vascular fragility. The additional component of the observed vascular fragility in this patient would be her recent drug rash. Given that the petechial rash appeared predominantly within the borders of the fading urticarial lesions, it is likely that the capillary dilation and local inflammation of the urticaria played a role in the susceptibility of the local vasculature to rupture. The

decreased capillary resilience due to long-term steroid use coupled with the increased capillary pressure secondary to the application of the tourniquet caused the dilated capillaries in the areas affected by the drug rash to rupture which resulted in the petechial/purpuric rash. The definitive diagnosis of acute capillary rupture syndrome (or RLP) is dependent on histopathology. Histopathological analysis enables the clinician to rule out alternative diagnoses. That being said, in this particular case, a cutaneous biopsy for histopathological analysis could not be justified given that the patient was asymptomatic (aside from the rash), the clinical suspicion for RLP was high, and that the histopathology was unlikely to affect management.

Patient's perspective

"Since being diagnosed with Still's disease as a young adult, my experience as a patient has been that there are two categories of coping and adjustment. The first category involves coping with the diagnosis itself—physically and psychologically. This involved adjusting to life with my diagnosis by working with my physicians to become more knowledgeable about its symptoms (eg, nocturnal fevers; non-itchy, lacey, salmon-coloured rash on my trunk; hot joint inflammation; sore throat) in order to better manage my health. This is important in situations like this recent one in which I was admitted to the hospital. When I came to the Emergency Department, I was able to discern that the particular rash I was experiencing was not consistent with the rash I experience during a Still's flair up. The presenting rash was a different colour, extremely itchy, appeared on all parts of my body as opposed to just my trunk, and I had no joint swelling or fever. I was able to communicate this information effectively to my doctors, who in turn were able to more quickly isolate and diagnose the real problem—a severe allergic reaction to some antibiotics I had been taking. In this situation, I feel like I was an effective advocate for my health.

The second category of adjustment I've had to make—which relates to this experience in particular—is becoming knowledgeable and managing the side effects of the maintenance medication I take to treat the Still's. Although I am thrilled to report that I have been in remission for over 2 years now, I take several medications to maintain this—anakinra, prednisone, hydroxychloroquine, folic acid and until recently methotrexate. While I'm aware of the more major potentially harmful side effects of these medications that I should be monitoring for (eg, hair loss; changes to my central vision; increased susceptibility to infection), my overall knowledge of the more general potential side effects of these medications remains limited. Before my experience with Rumpel-Leede phenomenon, I had no idea that some of the medications I take could lead to increased capillary fragility. As a patient, obviously I can appreciate the need for my physicians to prioritise in making me more aware of the more potentially dangerous side effects of these medications. From a patient perspective, however, I think this experience illustrates that there is something to be said for increasing patient knowledge about the more general side effects certain medications can have (such as capillary fragility); not simply the worst-case scenarios. As someone living with Still's, I believe that the most effective care is delivered when my physicians work with me to increase my overall knowledge as a patient, so that I have the skills necessary to be an effective advocate for my health—particularly in times of emergency."

Figure 3 The patient's arm at her 1-week follow-up appointment.

Learning points

- ▶ Long-term steroid use in patients with adult-onset Still's disease (AOSD) may induce a state of capillary fragility thereby predisposing those patients to Rumpel-Leede phenomenon (RLP).
- ▶ Patients who experience RLP should be reassured that, although the rash can appear serious, it will resolve spontaneously within 1–2 weeks with no lasting sequelae.
- ▶ Patients being treated for AOSD should be made aware that RLP is a potential complication of circumferential compression, as occurs with the application of a tourniquet or blood pressure cuff.
- ▶ The treatment of AOSD may induce a state of vascular fragility and therefore patients with AOSD should follow closely with their rheumatologists to minimise the complications of their disease, including RLP.

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Contributors JTL is the primary guarantor and primary author who identified and managed the case, and also performed planning and literature review. CM is the coauthor who performed editing and literature review.

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