

Patients with Lupus Anticoagulant/Antiphospholipid Syndrome and Bone Marrow Procedure: How Risky is it?

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ABSTRACT

Bone marrow (BM) examination is now considered a standard procedure for hematological diagnosis. It is usually harmless and without serious complications. We describe a patient with lupus anticoagulant who underwent a bone marrow aspiration and solid core biopsy for the investigation of possible myeloproliferative disorder (MPD). She developed considerable bleeding following the procedure. This incident highlights the importance of proper risk evaluation in patients with this syndrome prior to surgical manipulation.

Keywords: Lupus Anticoagulant; Antiphospholipid Antibody Syndrome; Bone Marrow Procedure

Abbreviations: LA: Lupus Anticoagulant; APS: Antiphospholipid Syndrome; APTT: Activated Partial Thromboplastin Time; PT: Prothrombin Time; DRVVT: The Diluted Russel's Viper Venom Time; EACA: Epsilon-Aminocaproic Acid

Introduction

Bone marrow examination using a bone marrow aspiration and a solid core biopsy needle has become a standard procedure for the evaluation, diagnosis, and management of patients with various hematological as well as non-hematological disorders [1]. In most instances (particularly when the provider is familiar with the technique) the procedure is usually harmless and without any significant complications. However, there are conditions when such a procedure can pose significant risk and morbidity in the form of extensive bruising or bleeding [1]. Although unappreciated patients with lupus anticoagulant (LA) and antiphospholipid syndrome (APS), also called antiphospholipid antibody syndrome may present one such complication. It is known that the presence of lupus anticoagulant is not only associated with an elevated risk of venous and arterial thrombosis but can also present serious bleeding complications. We describe such a patient with LA/APS who underwent a bone marrow procedure for the investigation of MPD and who developed prolonged bleeding from the site of the puncture for several days following the procedure.

Case Report

The patient is a 68-year-old white female who was referred to our clinic for the evaluation and management of leukocytosis, anemia, and thrombocytosis. Her past medical history was significant for anxiety, non-oxygen dependent COPD, hypothyroidism, hypertension, chronic back pain, neck pain with numbness in both hands, necrotizing fasciitis, varicose veins, superficial venous thrombosis left leg, and obesity. On physical examination, the patient was noted to be anemic but she was not in acute distress. There was no jaundice, cyanosis, or edema. Her abdomen was soft and non-tender. Bowel sounds were heard. The liver, spleen, and kidneys were not palpable. There was no palpable lymphadenopathy. Heart sounds were normal. The chest was clear to auscultation and her vital signs were stable. There were prominent bilateral lower extremity varicose veins. Laboratory investigations revealed WBC 11.6 x 10⁹/L, hemoglobin 6.4 g/dL with normal MCV (85.6 fl), but slightly low MCH (23.6 pg), and a slightly raised platelet count of 473 x 10⁹/L. The differential counts were normal. Her complete metabolic profile was normal. Her serum iron was low at 16 mcg/dl, iron-binding capacity was high at 476 mcg/dl, ferritin

was low at 6 ng/ml, and % transferrin saturation were low at 3%. Folic acid level was low at 5.7 ng/ml but B12 was slightly low at 154 pg/ml. Her thyroid function tests were normal except T3-triiodothyronine-total was slightly low at 69 ng/dl. Her activated partial thromboplastin time (APTT) was prolonged at 91.0 sec [normal value (NV) 25.0-34.0 sec].

But prothrombin time (PT) was normal at 12.7 sec (NV:11.0-15.0 sec) and INR was normal at 0.95. Evaluation of the clotting factors revealed slightly high levels of factor IX activity at 172 % (NV: 60-160%) and von Willebrand factor antigen at 259% (NV: 50-160%). A 1:1 dilution of patient plasma with normal plasma did not correct APTT

and the findings were compatible with circulating anticoagulant (probably lupus) involving intrinsic pathways. Thrombin time was 16 sec (NV: 12-19). The diluted Russel's viper venom time (dRVVT) was positive. The anticardiolipin antibodies IgG and IGM were positive. In this case, the findings of markedly prolonged APTT along with positive dRVVT and anticardiolipin antibody results were suggestive of LA/APS. In the absence of any previous history of bleeding manifestations, the patient underwent a bone marrow examination to elucidate the cause of her anemia with leukocytosis and thrombocytosis. However, following the bone marrow procedure the patient continued to bleed from the puncture site (Figure 1) and required hospitalization to control her bleeding.

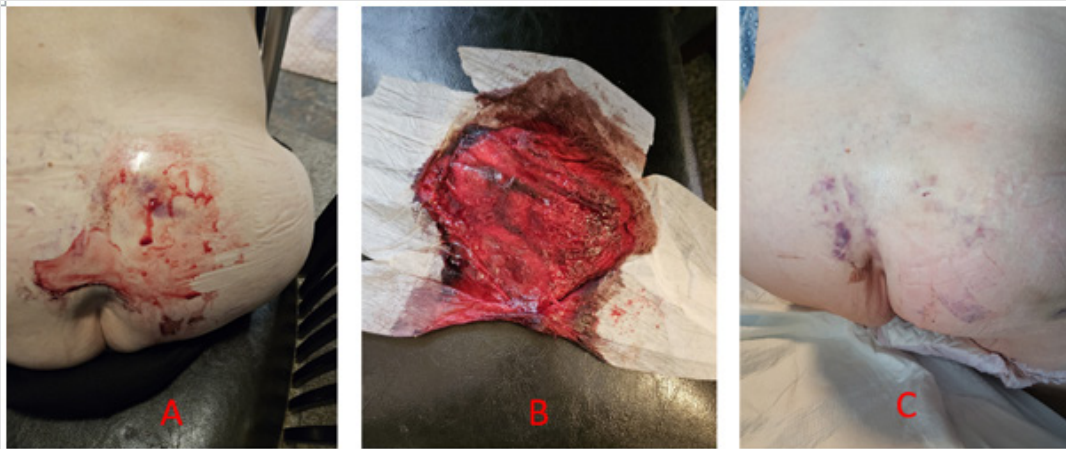


Figure 1. A - site of puncture, a day after the BM aspiration and biopsy procedure showing spurting of blood from the wound, B -gauze dressing (after overnight) showing the dressing soaked with blood, C- site of puncture seven days post-procedure showing residual bruise but no bleeding

Discussion

Lupus anticoagulant (LA) is an antiphospholipid antibody that causes prolonged in vitro coagulation times. This is a rare disease and is usually associated with a hypercoagulable state together with thromboembolic events [2]. A bleeding diathesis is a rare manifestation of lupus anticoagulant [3] and may be seen in surgical procedures or some type of hemostatic challenge (eg, dental extraction, trauma) [4] It has never been reported following a bone marrow procedure. We, report for the first time a patient with the presence of lupus anticoagulant who underwent a bone marrow procedure for the investigation of possible MPD who developed considerable bleeding following the procedure, required hospitalization and treatment with high doses of steroid, blood transfusion, fresh frozen plasma and Epsilon-aminocaproic acid (EACA) to stabilize her condition to stop the bleeding. Hematologic manifestations of LA/APS are relatively frequent and range from mild to severe. All three blood cell lines (red cells, white cells, and platelets) can be affected. Our patient presented with anemia, leukocytosis, and thrombocytosis (suggestive of MPD) for which she underwent a bone marrow examination.

Although the bone marrow procedure was uneventful, however, following the procedure she continued to bleed from the site and needed hospitalization to manage her bleeding. The case we presented here highlights the significance of adequate evaluation and treatment of patients with LA/APS before any surgical intervention. Although the most common clinical presentation of patients with LA/APS is arterial or venous thromboembolism [5] bleeding/hemorrhage may occur which is usually attributable to an associated thrombocytopenia, the platelet dysfunction, a prothrombin deficiency, or other underlying coagulopathies [6,7]. Our patient although carrying a diagnosis of LA/APS antibody syndrome did not have other hemostasis abnormalities. The bleeding complications although rare must be taken into account with LA/APS. Most of these cases usually respond well to administering prednisone treatment [3]. With hindsight, the prolonged bleeding following the bone marrow procedure in our patient was most likely due to her LA/APS and this complication may have been avoided if she had been treated with steroid to control her disease.

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