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Case Report

Spontaneous bleeding in systemic lupus erythematosus: Endovascular treatment of two rare cases

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ABSTRACT

Among the multiple clinical manifestations of systemic lupus erythematosus, spontaneous bleedings are rare but clinically important events. They could be potentially fatal, if not promptly treated.

The appropriate diagnosis, followed by the timely treatment of these rare clinical presentations, is essential to prevent their lethal consequences.

The purpose of this paper is to describe the diagnostic features and the endovascular treatment of 2 cases of spontaneous bleeding—respectively occurred in a 42-year-old woman with abdominal pain and melena, and in a 33-year-old woman with an extensive and painful hematoma in the left axillary region.

The timely endovascular treatment—performed by a minimally invasive approach of super-selective percutaneous embolization—has allowed an immediate clinical improvement, avoiding major surgery.

Background

Systemic lupus erythematosus (SLE) is a chronic, multisystem inflammatory disease of unknown etiology, characterized by development of cytological tissue due to deposition of circulating antibodies and immune complexes. Potentially, all organs may be affected; for this reason, clinical presentations of this pathology may be multiple and variable. Sponta-

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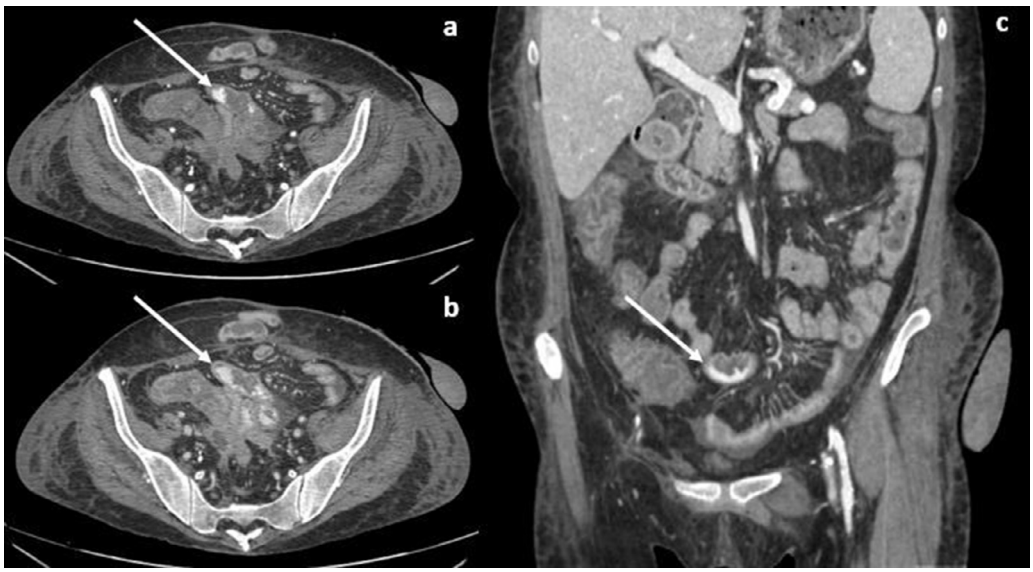

Fig. 1 – 42 years-old woman with SLE. Axial CT-angiography acquired during the arterial phase (a) and venous phases (b) showing active bleeding in the lumen of a small bowel loop (white arrow). The coronal reconstruction (c) shows the extravasation of iodinated contrast media on the inferior part of the small bowel lumen (with arrow).

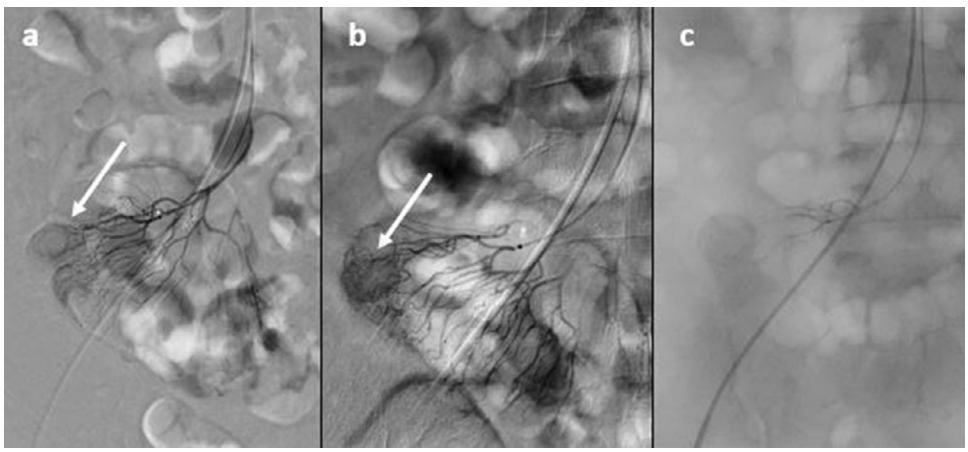

Fig. 2 – 42 years-old woman with SLE. Angiographic super-selective acquisitions (a,b) showing hyper - anarchic capillarization and early venous return (white arrows). The fluoroscopic control after embolization with gelfoam (c) shows the occlusion of the target vessels.

neous hemorrhagic manifestations are uncommon, and their underlying mechanisms have not been deeply elucidated [1].

Hereby we report 2 cases of spontaneous bleeding in patients with SLE, treated by endovascular super-selective embolization.

Cases presentation

Case 1

A 42-year-old woman was admitted to our emergency unit for abdominal pain; she had a remote pathological diagnosis

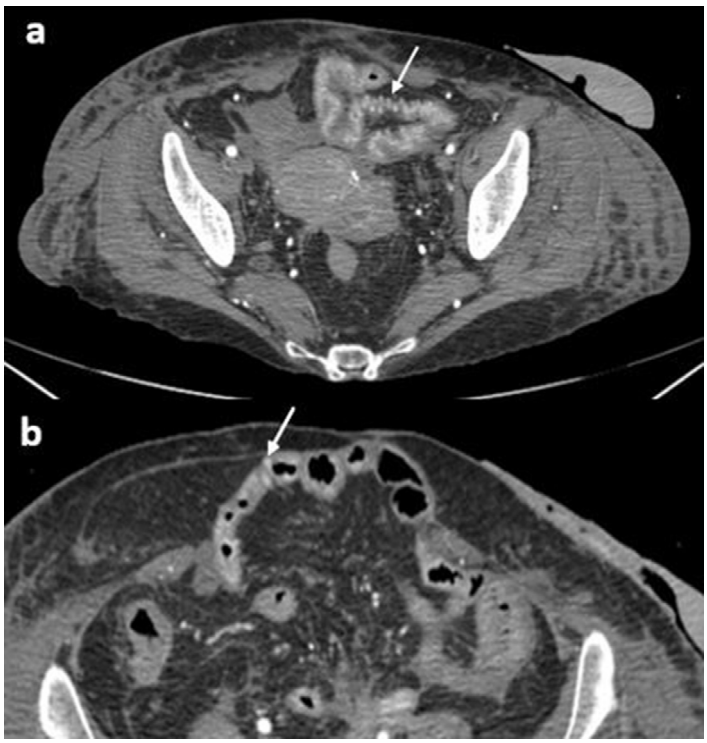

Fig. 3 – 42 years-old woman with SLE. Axial CT-angiography acquired during arterial phase showing diffuse hyperemia of the intestinal mucosa and sub-mucosal/intramural vascular ectasias, suggestive for angiodysplasia in the small bowel loops (white arrows).

of SLE from the age of 15, with chronic renal failure in dialytic treatment and a previous hemicolectomy for diverticulitis with a colostomy bag. During the hospitalization, anemia occurred with spontaneous bleeding from the colostomy. The CT-angiography (CTA) showed direct signs of active bleeding in the small intestine (Fig. 1).

The patient was hemodynamically stable. Thus she was urgently transferred to the angiographic suite where—after preparation of a sterile field and local anesthesia with lidocaine hydrochloride—a right common femoral arterial access was gained through a 5Fr vascular sheath, a Cobra C2-5Fr catheter was inserted on a hydrophilic wire (Terumo 180 cm), and advanced to the superior mesenteric artery (SMA). Arteriography of the SMA was performed, showing indirect signs of bleeding at the level of an ileal branch. Using a microcatheter (Boston Scientific Direxion Hi-Flo 0.027 inch x 155 cm) the feeding ileal arterial branch was superselectively catheterized: the arteriography revealed indirect signs of bleeding with anarchic- hypercapillarization and early venous return. Therefore, this arterial branch was embolized by using Spongostan. The final angiographic control showed a good morphological result with occlusion of the target vessel (Fig. 2). No peri or postprocedural complications occurred.

Four days later anemization occurred and a new CTA was performed. It revealed the presence of active contrast medium blush located on an ileal intestinal loop (distally to the previous treated region) with diffuse hyperemia of the intestinal mucosa and submucosal/intramural vascular ectasias, suggestive for angiodysplasia (Fig. 3).

Therefore, the patient was urgently transferred to the angiographic suite. A new SMA arteriography was performed demonstrating an active blush of a hypertrophic ileal branch of the SMA. Super-selective arteriography—perfomed with a coaxial microcatheter (Boston Scientific Direxion Hi-Flo 0.021 inch x 155 cm)–showed the presence of anarchic hypercapillarization at multiple levels, and confirmed the active blush. The involved vessel was embolized using Spongostan. The control arteriography of the SMA showed another arterial branch originating from a right hypotrophic colic artery that also supplied the blush. Therefore, also this branch was superselective catheterized using a microcatheter and embolized with Spongostan. The final arteriographic control showed no more blush (Fig. 4). After the treatment, anemia and vital's parameters improved and the patient was discharged some days later.

Case 2

A 33-year-old woman—with a clinical history of SLE in treatment with deltacortene and azathioprine, referring cough for about a month—was admitted to emergency due to sudden appearance of an extended hematoma with pain in the left axillary region and anemia. A CTA was acquired, showing active bleeding fed by branches from left lateral thoracic artery (Fig. 5).

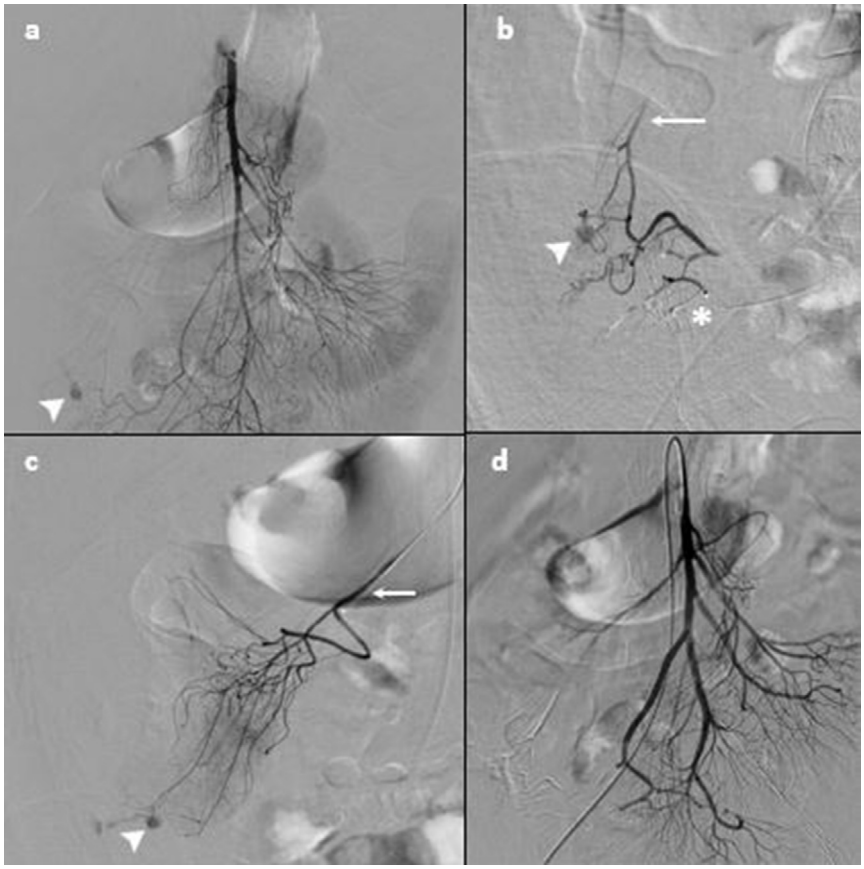

Fig. 4 – 42 years-old woman with SLE. Angiographic acquisition of the SMA (a) shows the active extravasation in an ileocolic branch (white arrowhead). The super-selective angiography of the ileocolic branches (b) by microcatheter (asterisk) demonstrates the contrast blush (white arrowhead) supplied by a collateral branch (white arrow). The subsequently super-selective angiography of the right colic artery (c) demonstrates the bleeding (white arrowhead) supplied also from a collateral colic branch (with arrow). After embolization of both target vessels with gelfoam, the final angiographic control from the SMA (d) shows no more extravasation.

The patient, hemodynamically stable, was transferred to the angiographic suite. With right common femoral artery access using a 5Fr sheath, the left subclavian artery was navigated using Bern shaped, 4Fr, 100 cm in length catheter and hydrophilic wire system (Terumo 180 cm). Diagnostic angiography documented direct and indirect signs of bleeding of 2 branches of the lateral thoracic artery. By means of a microcatheter (Cantata 2.5 Fr, 0.021 inch x 150 cm, Cook Medical) the aforementioned arterial branches were negotiated and embolized with Spongostan and detachable micro-coils (3 mm x 6 cm Interlock IDC-18, Boston Scientific), obtaining a good morphological result at the control arteriography (Figs. 6 and 7). The hematoma progressively resolved in the following days. Hemoglobin value and the others laboratoristic vital's parameters improved some days later, then the patient was discharged.

Discussion

SLE is a complex chronic, multisystem autoimmune disease due to the deposition of circulating antibodies and immune complexes that can affect any organ, resulting in extremely variable clinical features.

In SLE—the multisystemic involvement, the possibility of multiple scenarios of clinical presentations and the management of their complications—could make the diagnostic-therapeutic management of this pathology very complex.

In a recent systematic review, Lewis et al. [8] have reported an overall increasing time trend in both incidence and prevalence of SLE [9].

Therefore, it is essential to make a timely and correct diagnosis.

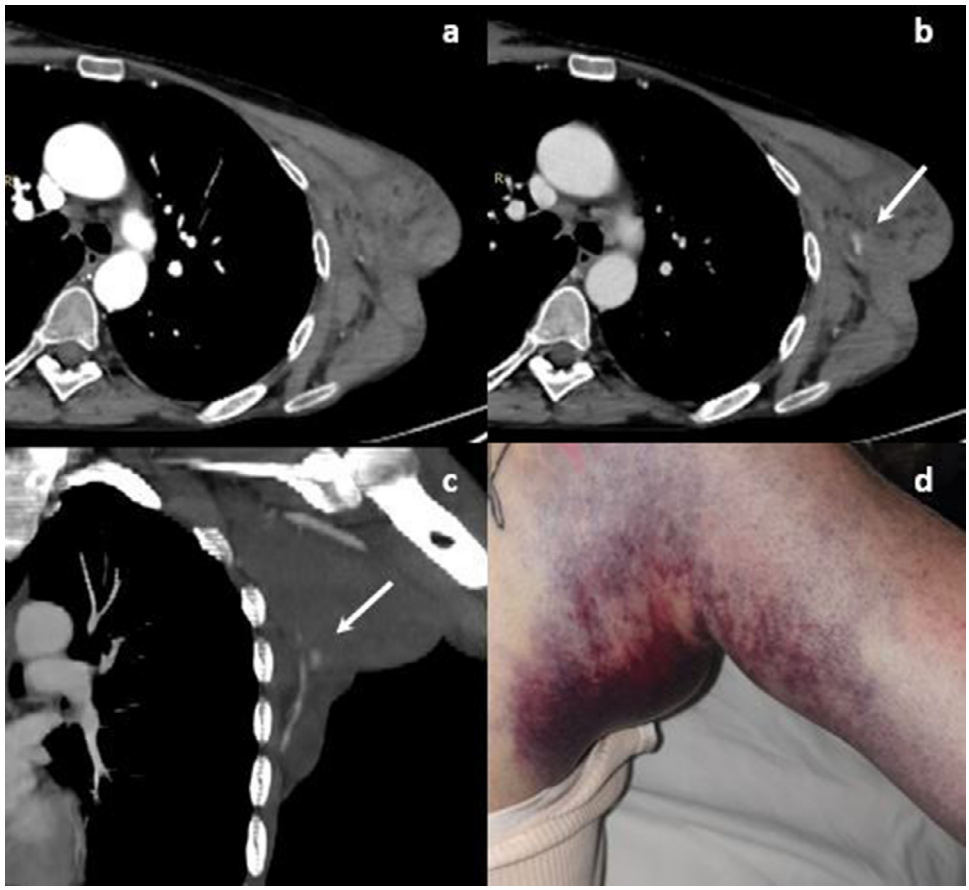

Fig. 5 – 33-year-old woman with SLE. Axial CT-angiography acquired during the arterial phase (a) and venous phases (b) showing active bleeding fed by branches from the left lateral thoracic artery (white arrow). The coronal reconstruction (c) confirmed the active extravasation (white arrow). Clinically, a visibly extended hematoma involved the left axillary region (d).

The new 2019 EULAR/ACR classification criteria for SLE include positive ANA at least once as obligatory entry criterion—followed by additive weighted criteria grouped in 7 clinical domains (constitutional, hematologic, neuropsychiatric, mucocutaneous, serosal, musculoskeletal, and renal) and 3 immunologic domains (antiphospholipid antibodies, complement proteins, and SLE-specific antibodies), weighted from 2 to 10. Patients accumulating ≥10 points are classified [10].

Among the various forms of intestinal involvement described in the literature, lupus mesenteric vasculitis (characterized by ischemic lesions) is certainly the most common, followed by others less common such as protein-dispersing enteropathy, intestinal pseudo-obstruction, acute pancreatitis, celiac disease, or inflammatory bowel disease. No specific autoantibody was associated with SLE-related forms of gastroenteropathy [6,11–13].

Typically, vasculitis in SLE patients affects vessels that are less than 100μ in diameter and is characterized by fibrinoid necrosis with marked wall thickening and minimal infiltration by inflammatory cells. This entity can occur in any organ or system, and can result in ischemia. In organs with terminal vascularization such as the bowel, vasculitis can compromise the blood supply to a segment of bowel, resulting in ischemia or hemorrhage into the bowel wall with subsequent perforation and peritonitis [12]. However, spontaneous hemorrhagic manifestations of SLE reported in the literature are rare, and may be potentially fatal if not promptly treated [1–5].

A proper diagnosis followed by the timely treatment of these rare clinical presentations is essential to prevent its potentially lethal consequences.

Among few cases reported in literature about spontaneous bleeding in SLE patients, the therapeutic management performed has mostly been based on medical or surgical treatment [2–4,6,7].

Only 1 case report has involved interventional radiology management so far [5].

These 2 cases have demonstrated a successful endovascular management of rare forms of spontaneous bleeding in patients with SLE, performing a minimally invasive approach of super-selective percutaneous embolization.

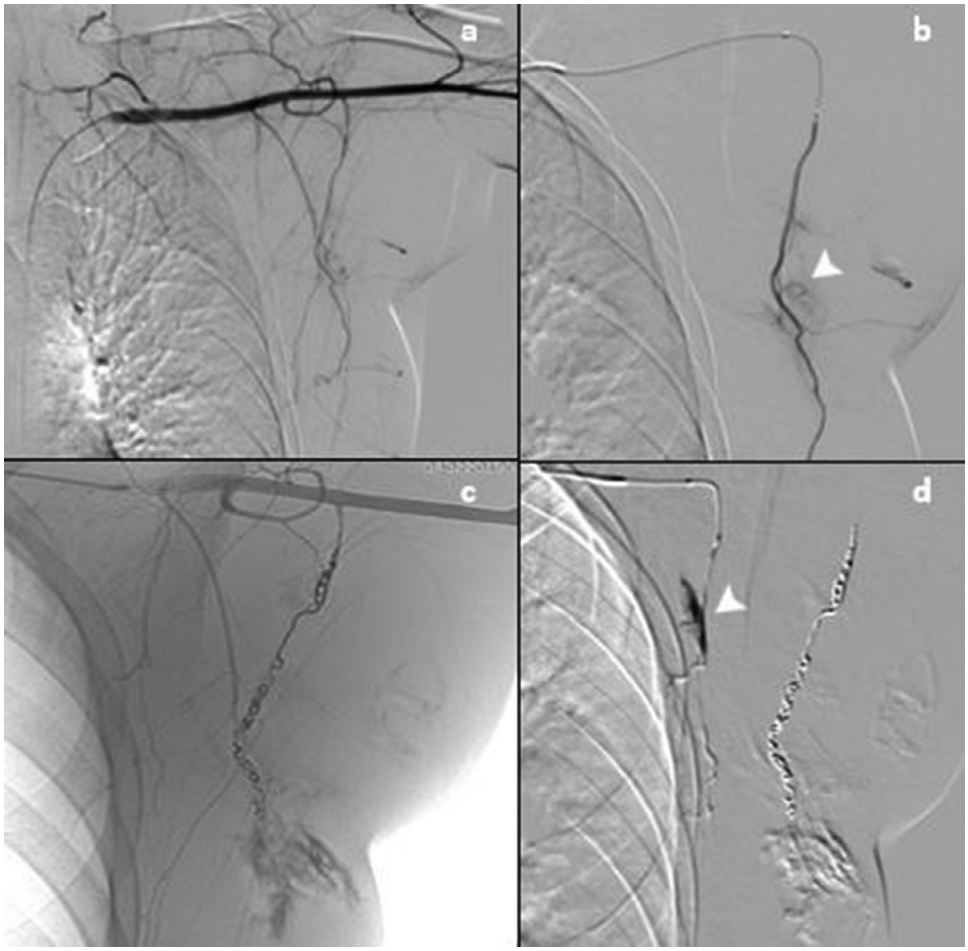

Fig. 6 – 33-year-old woman with SLE. Angiographic acquisition of the left subclavian artery (a) shows direct and indirect signs of bleeding of two branches of the lateral thoracic artery (a). The most lateral branch (b) was super-selective catheterized (b) to confirm the extravasation of contrast media (white arrowhead). The embolization was performed with 3 mm x 6 cm detachable micro-coils (c). Then, the super-selective angiography from the most lateral branch (d) confirmed the bleeding also from this vessel (white arrowhead).

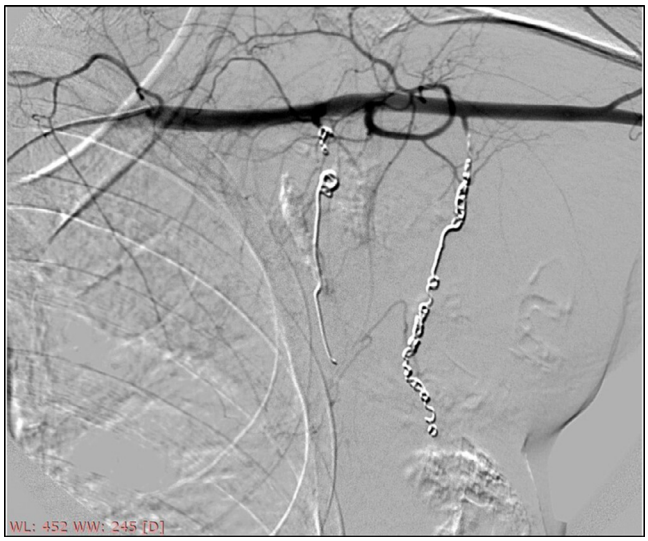

Fig. 7 – 33-year-old woman with SLE. The final angiographic control from the left subclavian artery shows the complete occlusion by detachable micro-coils of the target vessels.

The timely endovascular treatment of percutaneous embolization has allowed an immediate clinical improvement through a minimally invasive approach, avoiding major surgery such as, in the first case, further intestinal resections.

Therefore, considering the multisystemic involvement of the disease, in case of spontaneous bleeding in SLE patient, interventional radiology management should be advisable when possible.

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