

Gamna-Gandy nodules of the spleen and asplenism in SLE: a novel association?

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DESCRIPTION

We present a case of a 53-year-old woman who presented to the emergency room with acute abdominal pain, fever and haemodynamic and respiratory instability and was admitted to the intensive care unit with fulminant septic shock with multiorgan failure. CT imaging of the abdomen showed no gross abnormalities, initial laboratory results are presented in [table 1](#).

Despite adequate resuscitation, broad-spectrum antibiotics and supportive care, following the International Guidelines of the Surviving Sepsis Campaign, she died within a couple of hours. The patient had a history of systemic lupus erythematosus (SLE) with arthralgia, positive antinuclear antibodies and persistently high anti-DNA antibodies, but without active disease and no immunosuppressive medication since years.

Leucocyte typing of the initial peripheral blood smear showed Howell-Jolly bodies and diplococci, indicating massive bacterial load and functional asplenism. Functional asplenism or hyposplenism is present in up to 5% of patients with SLE with or without disease activity before.¹ No thrombocytosis or antiphospholipid antibodies—that are suggested as clues to autosplenectomy—were found in our patient.²

Postmortem blood cultures became positive for *Streptococcus pneumoniae*. Pathology revealed perivascular infiltrates in the liver and Gamna-Gandy bodies (GGB) in a remarkably small spleen ([figure 1](#)).

The patient did not take medication or drugs that could explain this image of the liver. GGB or haemosiderotic nodules are foci of haemosiderin deposition resulting from intrasplenic haemorrhage ([figure 2](#) and [figure 3](#)). Portal hypertension

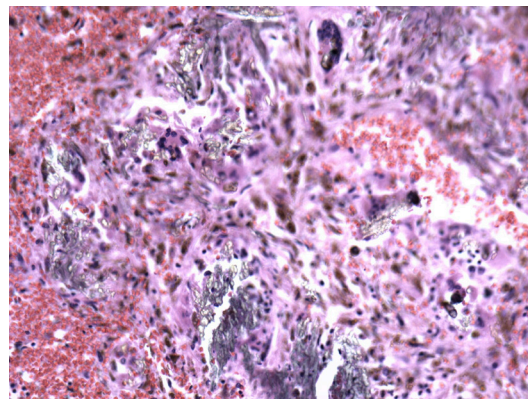


Figure 2 High-power view (HE ×200) of the Gamna-Gandy bodies. These rust-coloured foci contained fibrous tissue with haemosiderin deposits, and probably form due to scarring at sites of small perivascular haemorrhages.

is considered the primary cause of GGBs and their appearance is associated with sickle-cell anaemia, hereditary haemochromatosis, haemolytic anaemia, acquired haemosiderosis, paroxysmal nocturnal haemoglobinuria, portal or splenic vein thrombosis, leukaemia, lymphoma and repeated blood transfusion.³ However, these conditions known to be associated with GGB were absent in our patient.

Although the pathophysiological processes leading to hyposplenism in SLE remain largely unknown, it has been proposed that this is a result of silent infarction due to hyposplenism.⁴ As a consequence of inflammatory microvessel damage, splenic haemosiderin depositions may occur and remain visible, also after the resolution of vasculitis. Furthermore, despite the absence of antiphospholipid antibodies in this patient, splenic

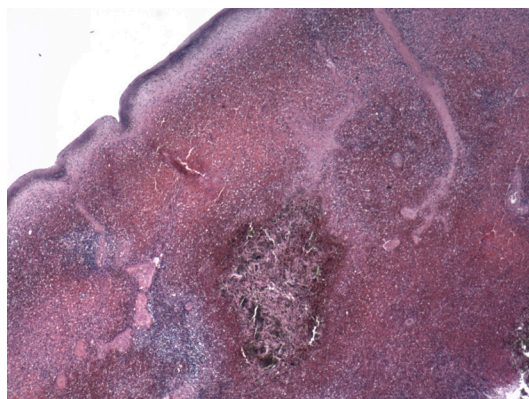


Figure 1 Microscopy of the spleen (HE ×25), revealing Gamna-Gandy nodules (centre).

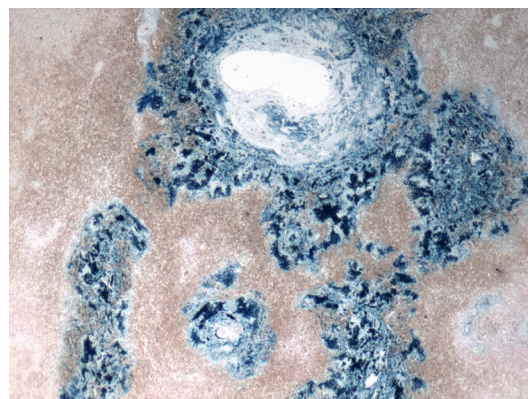


Figure 3 Prussian blue stain showing iron deposition (blue) in the Gamna-Gandy bodies.



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Table 1 Laboratory test results

Parameter	Value	Reference value	Unit
CRP	96	<10	mg/L
ESR	7	<20	mm/hour
Hb	8.0	7.0–9.2	mmol/L
Ht	0.40	0.32–0.44	L/L
MCV	95	82–89	fL
WBC	6.5	3.0–10.0	10 ⁹ /L
Platelet count	37	150–350	10 ⁹ /L
NB platelet count 10 days earlier: 318×10 ⁹ /L			
APTT	93	22–34	sec
PT	30.5	9.5–12.5	sec
Creatinine	411	44–80	µmol/L
Urea	17	2.5–6.4	mmol/L
Sodium	140	135–145	mmol/L
Potassium	4.0	3.5–5.0	mmol/L
Chloride	95	98–108	mmol/L
Glucose	0.6	3.0–7.0	mmol/L
Lactate	14.6	0.5–2.2	mmol/L
AP	118	<98	U/L
Gamma-GT	78	<38	U/L
ALAT	124	<34	U/L
ASAT	329	<31	U/L
LDH	1748	<247	U/L
Amylase	97	<107	U/L
CK	1269	<145	U/L
Arterial blood gas			
pH	7.24	7.35–7.45	
pCO ₂	2.6	4.6–6.4	kPa
pO ₂	17.2	10.0–13.3	kPa
HCO ₃	8.2	22.0–29.0	mmol/L
BE	–17.1	–3.0–3.0	mmol/L
sO ₂	98	95–98	%

ALAT, alanine aminotransferase; AP, alkaline phosphatase; APPT, activated partial thromboplastin time; ASAT, aspartate aminotransferase; BE, base excess; CK, creatine kinase; CRP, C reactive protein; Gamma-GT, gamma-glutamyl transferase; Hb, haemoglobin; Ht, haematocrit; LDH, lactate dehydrogenase; MCV, mean corpuscular volume; pCO₂, partial pressure of carbon dioxide; pO₂, partial pressure of oxygen; sO₂, oxygen saturation; PT, prothrombin time; WBC, white blood cell.

microangiopathy and microthrombosis associated with SLE may have contributed to ‘leaky’ splenic microvasculature, resulting in GGBs and partial autosplenectomy as described above.

In conclusion, this case once again illustrates the association of SLE with asplenia and the fulminant course of pneumococcal sepsis in those patients,⁵ despite disease inactivity for years. Moreover, GGB were found, which could not be related to known associated diseases. As far as we know, this is the first case describing GGB in a patient with SLE and functional asplenia.

Learning points

- ▶ Functional asplenia or hyposplenia is present in a small but significant part of patients with systemic lupus erythematosus (SLE), regardless of disease activity and use of immunosuppressive medication. Diagnostic clues indicating a susceptibility to splenic hypofunction or dysfunction remain uncertain, making it impossible to predict which patients will develop asplenia.
- ▶ Taking into account the fulminant course of pneumococcal infection in patients with SLE and asplenia, prophylactic vaccination should be considered in every patient with SLE.^{4,5}
- ▶ This is the first case that describes GGB in a patient with SLE and functional asplenia.

Contributors LEMH and AJP: contributed equally to the writing of this manuscript and were personally involved with the case. AM: provided the pathology images and wrote the figure captions. ML: shared his expertise on the subject in trying to explain the findings in this particular setting. AM and ML: reviewed the draft text.

Competing interests None declared.

Patient consent Obtained.

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