CASE REPORT

Recurrent syncope in systemic lupus erythematosus: a hidden cause in abdomen

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SUMMARY

Systemic inflammatory rheumatic diseases have shown an increase in frequency of internal malignancies, predominantly lymphoproliferative disorders. Occurrence of solid organ tumours is exceedingly rare. It is even rarer for it to manifest as recurrent syncope. We report a 55-year-old woman with systemic lupus erythematosus, who later developed episodes of syncope and dizziness along with diaphoresis and palpitations. She also had associated abdominal pain and vomiting. Imaging revealed a gall bladder (GB) mass with hepatic extension, which was histologically consistent with adenocarcinoma of the GB. Subsequently she succumbed to death during chemotherapy.

BACKGROUND

Collagen vascular disorders have been associated with a high risk of internal malignancies. The association could be incidental, secondary to persistent inflammation, paraneoplastic or treatment-related. We report a rarer association of carcinoma of the gall bladder (GB) in a case of systemic lupus erythematosus (SLE) with an unusual clinical presentation of recurrent syncope.

CASE PRESENTATION

A 55-year-old woman presented for evaluation of photosensitivity, episodes of painless oral ulcers, polyarthralgia, Raynaud's phenomenon and progressive dysphagia to solid foods that were present for 3 years and significant weight loss during the past 1 year. Her medical history was unremarkable except for active tobacco smoking for the past 10 years.

Physical examination revealed multiple small depressed and atrophic plaques over the tip of nose, chin, bilateral ear concha, upper back and bilateral soles (figure 1A–C). She had non-scarring alopecia with short broken and lusterless hairs, digital pitted scars, livedoid changes (figure 1D) and pseudo-ainhum over the second right toe.

INVESTIGATIONS

She had a positive antinuclear antibody with a titre of 1:320 (speckled) along with anticardiolipin antibodies (38.7; n=0–12). Anti-dsDNA (16 IU/mL (0–50)), Rheumatoid arthritis factor and direct Coomb's test were negative. Anti-Scl-70 and anticentromere antibodies levels could not be measured. Barium swallow revealed dilatation of the distal oesophagus with motility disorder, and

high-resolution CT of the chest showed features of non-specific interstitial pneumonia pattern along with dilated oesophagus (figure 2A). Other investigations including haemogram, metabolic panel and urinalysis were unremarkable. Baseline ECG was normal and echocardiography showed trivial MR. Holter ECG showed normal sinus rhythm with heart rate 59–141 beats per minute along with occasional supraventricular and ventricular ectopics. Routine autonomic function test revealed a fall of 40 mm Hg in systolic blood pressure by head up tilt test. Peripheral nerve conduction studies were normal. With these clinical history and investigations, a final diagnosis of SLE was made.

Shortly after admission, she had multiple episodes of syncope and dizziness along with diaphoresis and palpitation while standing and walking that resolved on lying down. There were associated episodes of acute abdominal pain and vomiting. In view of these, ultrasonography of the abdomen was carried out that revealed a GB mass infiltrating into the adjacent liver parenchyma with focal thickening of its wall and cytology revealed atypical cells. Multi-phase contrast-enhanced CT also revealed focal wall thickening of fundus of GB with infiltration in adjacent segment V of liver along with pericholecystic fluid collection suggestive of localised rupture of GB (figure 2B–D). With these features she was diagnosed to have stage 4 carcinoma of GB with liver metastasis.

OUTCOME AND FOLLOW-UP

She was transferred to oncology department for further management, and during the follow-up after chemotherapy, the repeat CECT showed progressive disease with multiple metastases in the liver (figure 2E–F). Later she succumbed to death.

DISCUSSION

Collagen vascular disorders such as SLE and systemic sclerosis have been associated with an increased risk of cancer.^{1–3} In SLE, the occurrence of haematological malignancies, in particular Hodgkin's lymphoma, is increased about three-fold. Threefold. Of these, diffuse large B cell lymphoma is the most frequent tumour. Patients with SLE are also at an increased risk of developing lung (standard incidence ratio (SIR) 1.30; 95% CI 1.04 to 1.60), cervical (SIR 1.27; 95% CI 0.78 to 1.93) and head and neck cancers (SIR 2.16; 95% CI 1.13 to 4.13).⁴ The occurrence of GB cancer is exceedingly rare, and in a large

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Figure 1 (A–C) Atrophic plaques and scarring over the face, lower back and soles. (D) Livedoid changes on the medial aspect of thigh.

series of 96 cases of SLE, only 5 had solid tumours including a case of carcinoma of GB.¹

Cardiac involvement is the most common cause of syncope in collagen vascular disorders mostly resulting from conduction disturbances, arrhythmias, myocardial and valvular involvement or autonomic neuropathy.⁵ In our case, ECG, echocardiography and Holter monitoring were normal and hence cardiac causes

were excluded. The syncope in our case was also quite unlikely to be due to collagen vascular disorder -related autonomic neuropathy as it was recent in onset. Alternatively it may be due to reflex-mediated neurocardiogenic syncope resulting from the GB tumour, its distension and rupture. GB is richly supplied with sympathetic and parasympathetic innervation. It has been postulated that GB distension secondary to inflammation may

Figure 2 (A) Axial CT image in lung window shows ground glass opacity in basal segment of both lower lobes with interlobar septal thickening (arrows) suggesting interstitial lung disease and dilated distal oesophagus (vertical arrow). (B) Contrast-enhanced CT of abdomen shows focal wall thickening of fundus of gall bladder (GB) with infiltration in adjacent segment V of liver (arrows), suggestive of malignancy. (C) (Coronal) and (D) (axial) images showing pericholecystic fluid collection suggestive of localised rupture of GB (arrows). (E and F) Follow-up CT done after chemotherapy shows increase in the extent of primary GB mass (white arrow) and few non-contiguous hypodense metastatic lesions in liver (black arrow).

result in increased reflex sympathetic discharge in the spinothalamic tract, which in turn stimulates the baroreceptor pathway and reflex increase in vagal tone, leading to bradycardia and syncope.⁶ In this case, the episodic syncope was also associated with recent history of significant weight loss, anorexia and severe abdominal pain, which further reiterates our hypothesis of GB-related syncope.

In conclusion, the purpose of reporting this case is to highlight the rarer association of carcinoma of the GB in SLE. Another interesting aspect of this report is recurrent episodes of syncope, which may be related to carcinoma of the GB itself. Hence in the context of ancillary features of malignancy such as anorexia, weight loss and abdominal pain in a patient with

SLE, a thorough screening of hepatobiliary system needs to be done.

Contributors TD, NB and GS evaluated and managed the case. TD, NB, GS and PN drafted the manuscript. All four authors finally approved the manuscript. PN provided radiological images for the manuscript.

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REFERENCES

1. Lopez Dupla M, Khamashta M, Pintado Garcia V, *et al.* Malignancy in systemic lupus erythematosus: a report of five cases in a series of 96 patients. *Lupus* 1993;2:377–80.
2. Lewis RB, Castor CW, Knisley RE, *et al.* Frequency of neoplasia in systemic lupus erythematosus and rheumatoid arthritis. *Arthritis Rheum* 1976;19:1256–60.
3. Tarr T, Györfy B, Szekanecz E, *et al.* Occurrence of malignancies in Hungarian patients with systemic lupus erythematosus: results from a single center. *Ann N Y Acad Sci* 2007;1108:76–82.
4. Tessier-Cloutier B, Clarke AE, Ramsey-Goldman R, *et al.* Systemic lupus erythematosus and malignancies. *Rheum Dis Clin North Am* 2014;40:497–506.
5. Prochaska MT, Bergl PA, Patel AR, *et al.* Atrioventricular heart block and Syncope coincident with diagnosis of systemic lupus erythematosus. *Can J Cardiol* 2013;29:1330.e5–1330.e7.
6. Vloka ME, Tamis JE, Steinberg JS. Neurally mediated Syncope in 2 patients with extracardiac disease. *Arch Intern Med* 1999;159:625–7.

Learning points

- Occurrence of hepatobiliary malignancy in systemic lupus erythematosus is extremely rare.
- It may be reflex mediated neurocardiogenic syncope resulting from the gall bladder tumour, its distension and rupture. Unexplainable recurrent syncope in such patients with constitutional features of anorexia, weight loss and abdominal pain should alert the clinician to look for hepatobiliary malignancy.

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