Case 17782

Eurorad

Large splenic mass in a young female

Published on 30.06.2022

DOI: 10.35100/eurorad/case.17782
ISSN: 1563-4086
Section: Abdominal imaging
Area of Interest: Spleen
Imaging Technique: CT
Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 27 years, female

Clinical History:

A 27-year-old female with Behçet's disease presented with two hours of severe non-radiating epigastric pain associated with breathlessness and nausea. Vitals were within normal limits, but patient could not tolerate physical examination. The possibility of pulmonary embolism and acute coronary syndrome was considered, and CT pulmonary angiogram (CTPA) was performed.

Imaging Findings:

CTPA was negative for pulmonary embolism. However, upper abdominal sections showed an enlarged spleen with a large hypodense lesion. Subsequent abdominal ultrasound revealed an enlarged spleen with a large, predominantly hyperechoic lesion occupying nearly the entire spleen with internal vascularity (Fig 1). MRI demonstrated a 10.6cm x 11.7cm x 14.7cm lesion hypointense to spleen on T1-weighted images. On T2-weighted images, the lesion was heterogeneously hyperintense with multiple tiny cystic areas. The lesion demonstrated some areas of diffusion restriction with no evidence of haemorrhagic content. Dynamic post-contrast MRI showed heterogeneous enhancement with multiple non-enhancing areas and few areas of delayed enhancement (Fig 2). Differential diagnosis of splenic haemangioendothelioma, haemangioma and angiosarcoma was considered. PET CT was conducted to further evaluate for primary or secondary tumours and showed no significant uptake in the splenic lesion and no other areas of increased activity (Fig 3).

Discussion:

The spleen is often considered as the "forgotten" and "mysterious" organ of the abdominal cavity. The space-occupying lesions of the spleen could be of different nature: congenital, inflammatory, traumatic, vascular, haematological and neoplastic diseases [1]. Splenic neoplastic lesions can be benign or malignant and there is considerable overlap in the imaging features. Lymphoma and metastasis are the common malignant lesions and common benign tumours include haemangioma, hamartoma and lymphangiomas [2].

Splenic haemangiomas are the most common benign tumour of the spleen, with an estimated incidence of 0.03-14% from a large autopsy study [3] occurring more frequently in male patients [4]. Splenic haemangiomas are thought to be vascular malformations of congenital origin arising from sinusoidal epithelium [5]. Splenic haemangioma is usually seen in adults between their third and fifth decades of life [6], however, rare cases in the paediatric population have also been reported [7].

Splenic haemangiomas can occur as solitary (more commonly) or multiple lesions and may represent a manifestation of systemic angiomatosis (associated with Klippel-Trénaunay-Weber syndrome and Beckwith-Wiedemann syndrome) [8]. Most splenic haemangiomas are small (<4cm in diameter) and asymptomatic, while large lesions (?4cm) are more likely to be symptomatic [3, 6]. Symptoms include pain and fullness over left upper quadrant, nausea and vomiting, and diarrhoea or constipation [4]. Physical examination may reveal a palpable mass in case of a large lesion and laboratory testing may reveal anaemia, thrombocytopenia and/or coagulopathy[9] . Large lesions also have a higher likelihood of spontaneous rupture with life-threatening haemorrhage[3, 6] . Therefore, splenic haemangiomas are important to identify and characterize for optimal patient management and outcome.

The appearance of splenic haemangiomas on imaging is similar to haemangiomas elsewhere in the body. Most haemangiomas are well-circumscribed, hypo- to isointense on T1-weighted images and hyperintense on T2-weighted images in relation to splenic parenchyma. The contrast-enhanced study can show centripetal enhancement in the early stages with progression to uniform enhancement in delayed images[1, 4, 5, 10] . However, splenic haemangiomas may demonstrate discrete mottled areas of heterogenous enhancement rather than the typical centripetal enhancement seen with typical liver haemangioma [9]. Well-defined peripheral nodules that coalesce over time seen in hepatic haemangiomas are not commonly seen with splenic haemangiomas. This is likely secondary to the difference in the vascular supply of the background organ rather than inherent differences between splenic and hepatic haemangiomas [11].

Splenic haemangiomas are slow-growing tumours and can be managed with observation in asymptomatic patients with small lesions. However, spontaneous rupture in 25% of cases has been reported [3, 4]. Therefore, large symptomatic lesions require treatment with splenectomy (more commonly) or embolization [6]. In the index case, splenectomy was performed and histopathology revealed haemangioma with no malignant features.

Teaching points

- Splenic haemangiomas are rare, slow-growing, benign vascular tumours
- Splenic haemangiomas are usually small, asymptomatic, and found incidentally on imaging
- MRI imaging features: hypo- to isointense on T1WI, hyperintense on T2WI, and heterogenous mottled enhancement on dynamic contrast images
- Small lesions (<4cm in diameter) can be managed with observation

Large lesions (?4cm in diameter) require treatment with splenectomy due to higher likelihood spontaneous rupture with intra-abdominal haemorrhage

Differential Diagnosis List: Splenic angiosarcoma , Splenic hamartoma , Splenic haemangioma, Splenic haemangioendothelioma , Lymphoma , Metastases

Final Diagnosis: Splenic haemangioma

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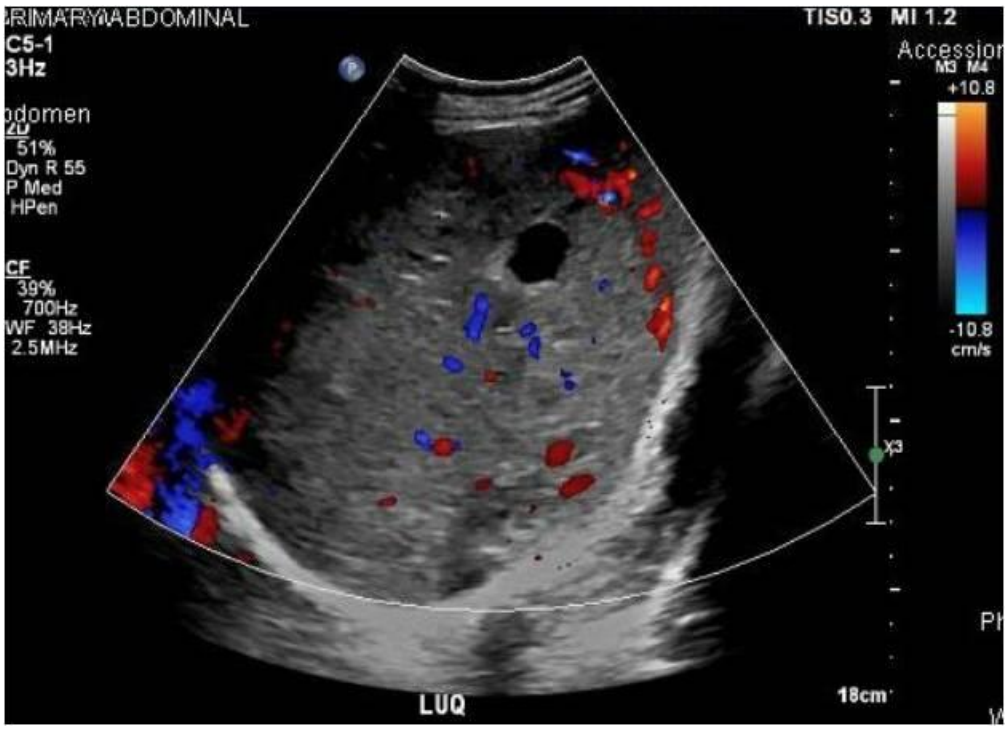
Figure 1

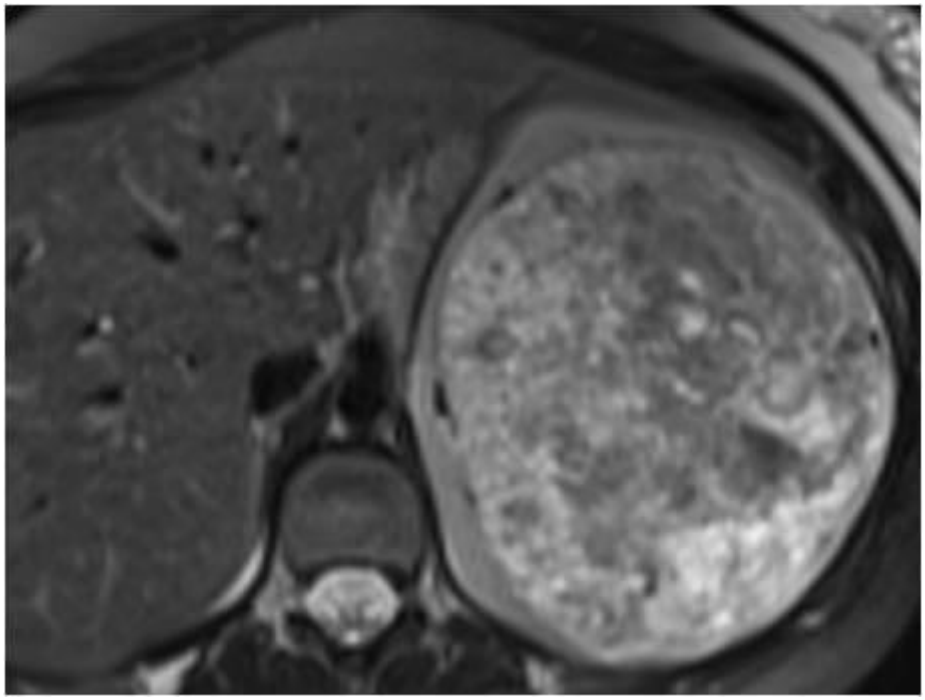
a



Description: Color doppler ultrasound shows internal vascularity within the large splenic lesion **Origin:**

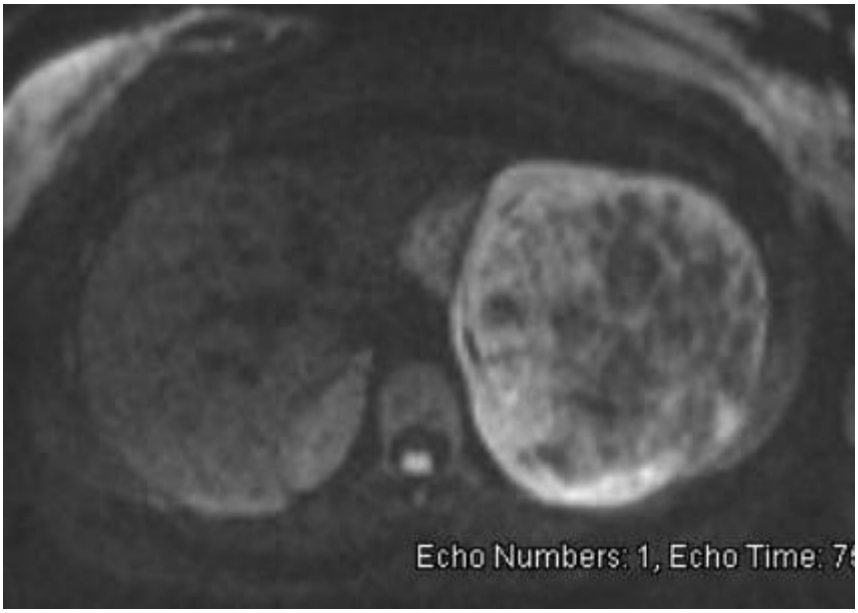
Figure 2

a



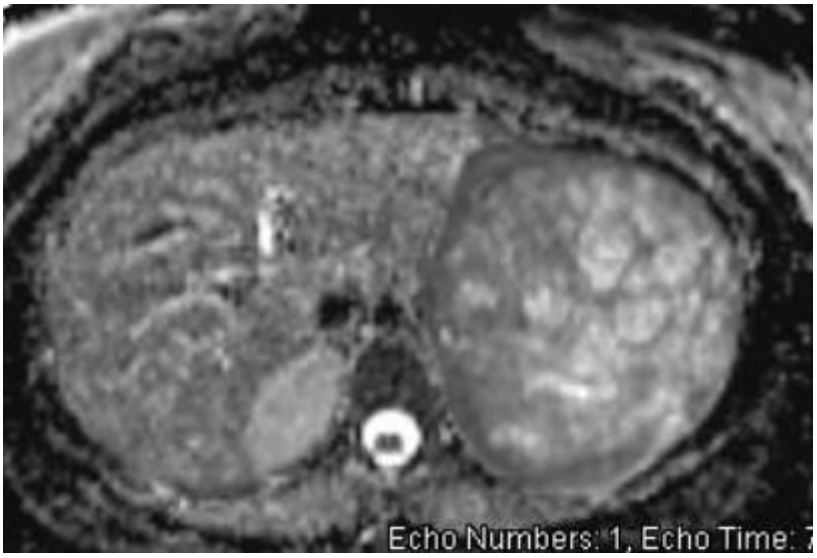
Description: Axial T2 image shows large heterogenous hyperintense lesion with multiple tiny cystic areas **Origin:** © Department of Clinical Imaging, Hamad Medical Corporation, Doha, Qatar, 2022

b



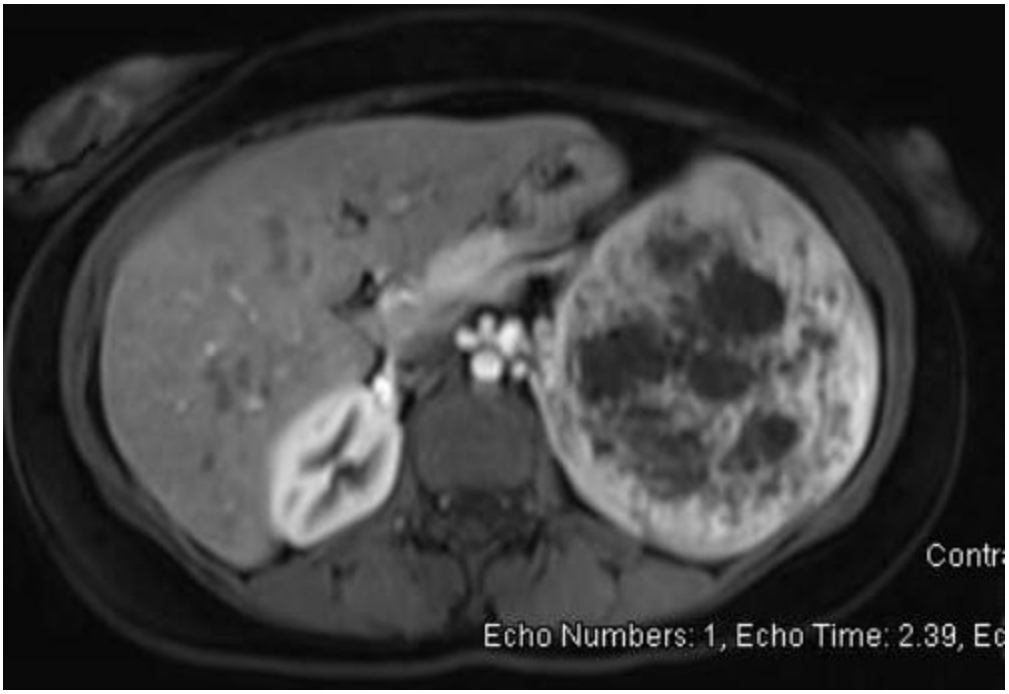
Description: Axial diffusion weighted image shows areas of diffusion restriction on high b-values **Origin:** © Department of Clinical Imaging, Hamad Medical Corporation, Doha, Qatar, 2022

c



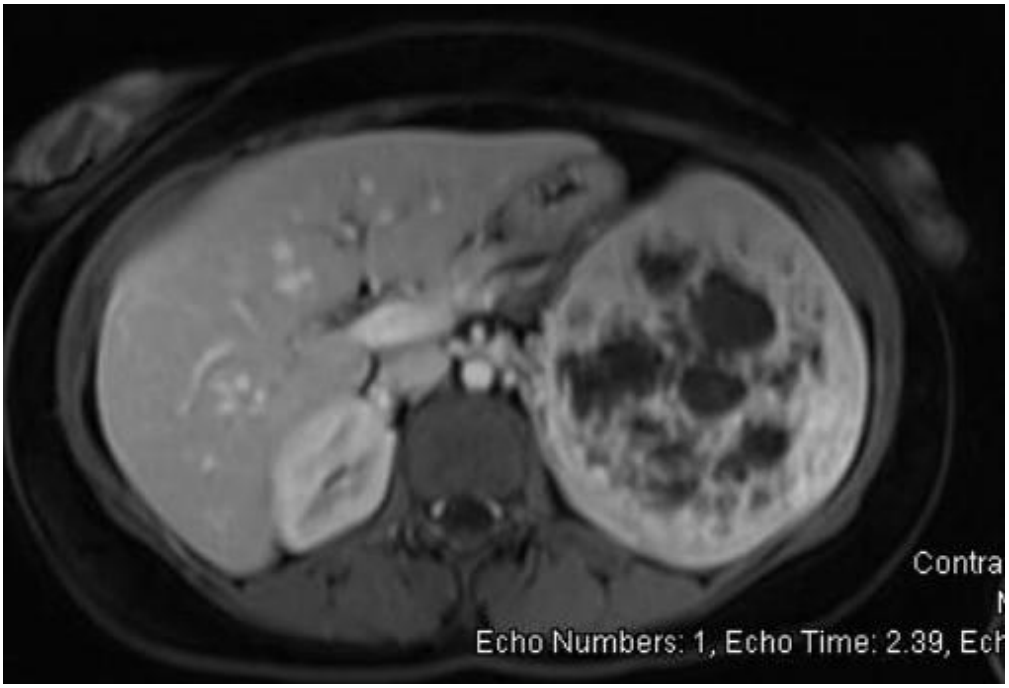
Description: Axial ADC map shows low signal in corresponding areas of high diffusion **Origin:** © Department of Clinical Imaging, Hamad Medical Corporation, Doha, Qatar, 2022

d



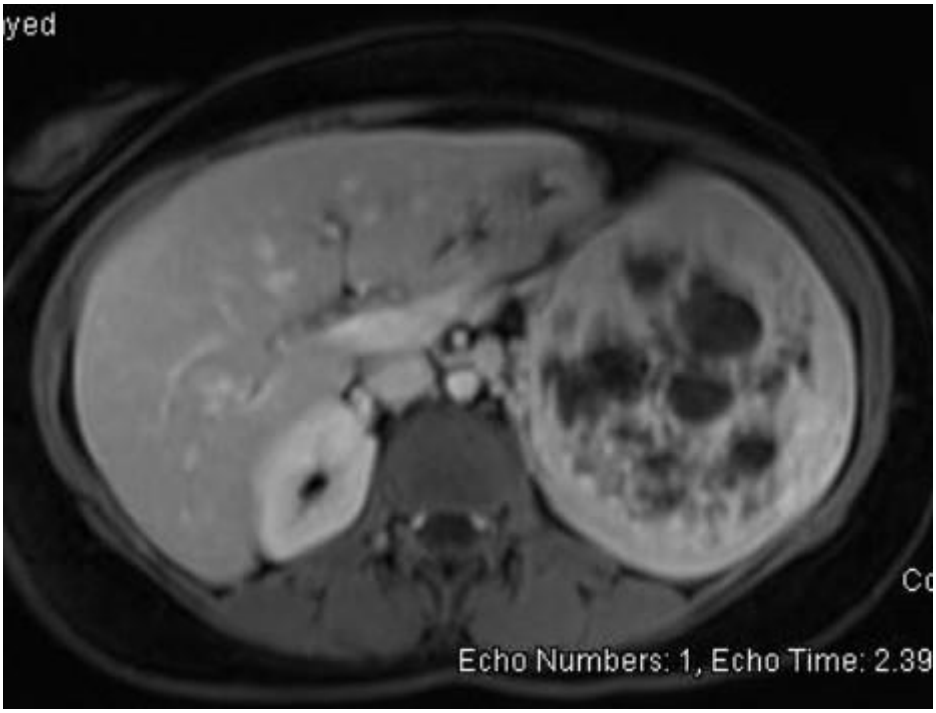
Description: Axial arterial phase postcontrast T1 image shows heterogenous enhancement with multiple areas of non-enhancement **Origin:** © Department of Clinical Imaging, Hamad Medical Corporation, Doha, Qatar, 2022

e



Description: Axial portal venous phase postcontrast T1 image shows heterogenous enhancement with internal areas of non-enhancement **Origin:** © Department of Clinical Imaging, Hamad Medical Corporation, Doha, Qatar, 2022

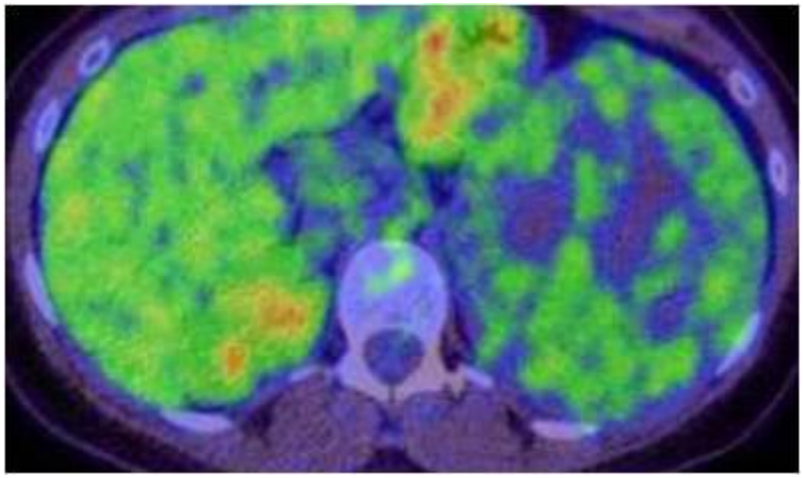
f



Description: Axial delayed phase postcontrast T1 image shows heterogenous enhancement with internal areas of non-enhancement and few areas of delayed enhancement **Origin:** © Department of Clinical Imaging, Hamad Medical Corporation, Doha, Qatar, 2022

Figure 3

a



Description: Axial PET CT image shows no significant uptake in the splenic lesion **Origin:** © Department of Clinical Imaging, Hamad Medical Corporation, Doha, Qatar, 2022