

Ectopic Splenic Tissue: the value of contrast enhanced ultrasound.

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Background

Two forms of ectopic splenic tissue are commonly recognised, splenosis and splenunculi (accessory spleens). Both are often discovered as incidental findings and commonly pose a diagnostic dilemma due to their ability to mimic more sinister pathology. As splenic tissues have a characteristic sonographic appearance, ultrasound is a valuable tool in making the diagnosis. The characteristic feature of ectopic splenic tissue with contrast enhanced ultrasound (CEUS) is persistent late phase enhancement due to microbubble sequestration. This will usually persist for 5-7 mins post injection - well after microbubbles have cleared from other normal and pathological tissues. Ectopic splenic tissue will also usually show an inhomogeneous early phase enhancement pattern ("zebra striped"). The CEUS behaviour is identical to normal spleen.

Splenosis

Splenosis is a form of ectopic spleen tissue caused by auto transplantation of native splenic tissue. Traumatic rupture and splenectomy are both recognised causes. It can occur both inside and outside of the peritoneal cavity, deposits being reported within the thorax, abdomen, pelvis, and subcutaneously. They can be distinguished from splenunculi due to location and their differing vascular supply.

Splenosis is usually asymptomatic and may be noted as an incidental finding during routine US, CT, MRI, or even during surgery. As with splenunculi the tissue is functional and thus may be beneficial for the immune system of the patient, reducing the risk of infection and maintaining normal immunological function.

Management of asymptomatic splenosis is therefore conservative. In rare cases, complications such as abdominal pain or obstruction, may occur, and indicate the need for resection of the ectopic tissue.

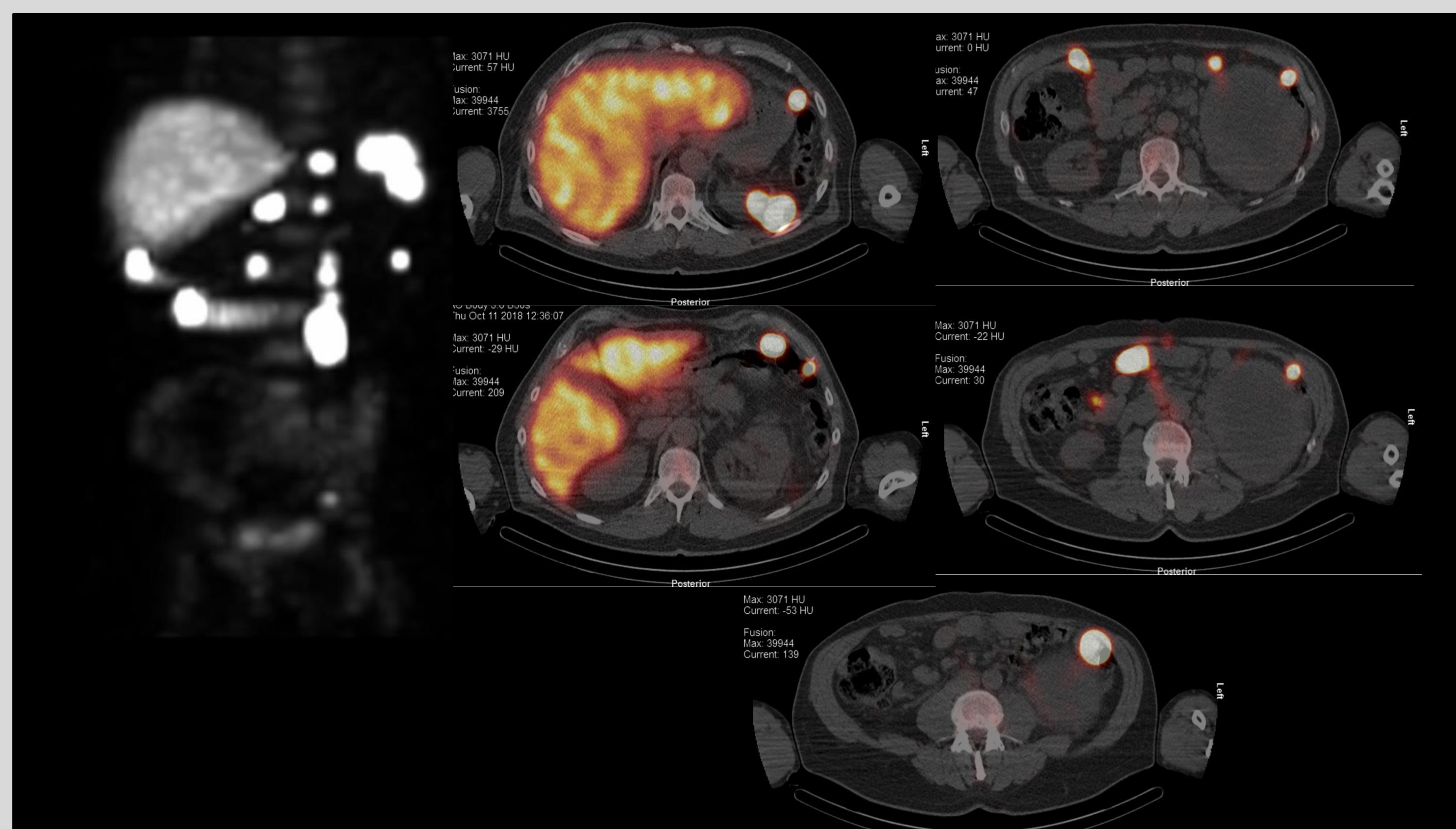
Issues are predominately related to misdiagnosis. The varying locations of deposits (often intravisceral), can lead to the misdiagnosis, most often of malignancy. This can lead to unnecessary investigations or even surgery, necessitating the need for a high index of suspicion in cases where previous splenic trauma is known or suspected.

Splenunculi (accessory spleens)

Accessory spleens consist of benign splenic tissue of congenital origin that arise due to incomplete fusion of splenic buds. Often small, rounded nodules, most are located around the splenic hilum or ligaments, around the pancreatic tail or left upper quadrant above the renal pedicle. They may also be found in sub diaphragmatic, pararenal and gastric locations. The majority occur as single nodules, however it is not infrequent for two or more nodules to be observed adjacent to each other. Size can be variable, ranging from mm's to cm's. Hypertrophy post splenectomy is a well recognised feature.

They are almost universally asymptomatic. Rare cases of torsion, rupture and bleed have been described, however as with splenosis morbidity is predominately related to their ability to mimic other masses and lymphadenopathy. In the absence of these complications, or other rare situations such as of lymphoma recurrence or hypersplenism no treatment is required.

Incidence is reported to be 10-30% at autopsy.



Widespread splenosis

These planar and SPECT CT images from a denatured red cells scan show a case of numerous deposits within the abdominal cavity all consistent with splenosis in a patient with a known renal tumour.

CEUS was not used to confirm the diagnosis in this case due to the number of lesions present.

Imaging in ectopic splenic tissue

The principles of imaging in ectopic splenic tissue apply to both splenosis and splenunculi. In all modalities they retain the same appearances (excepting location and size) and enhancement patterns as normal spleen.

Nuclear medicine tests such as Technetium (Tc-99m) sulfur colloid or Tc-99m heat-damaged red blood cell (RBC) scans have classically been the definitive test due to their high sensitivity and specificity.

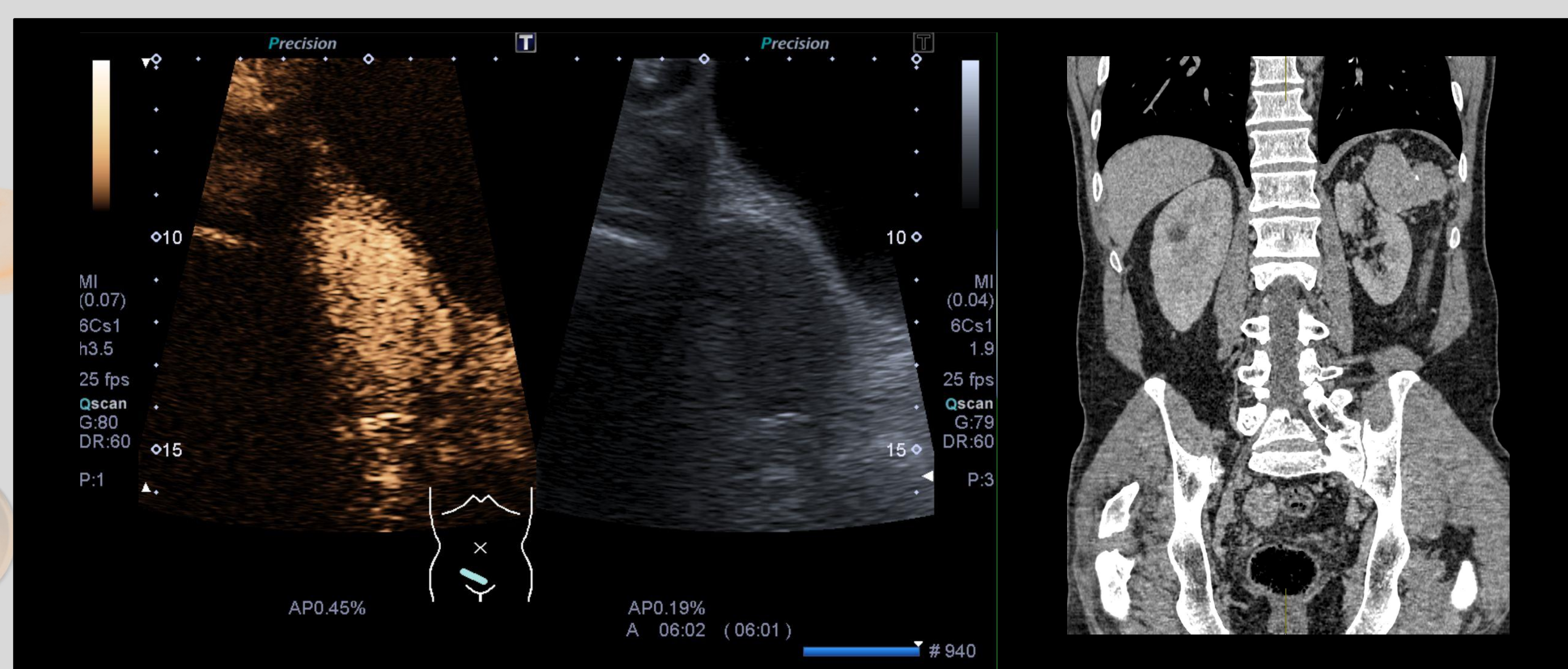
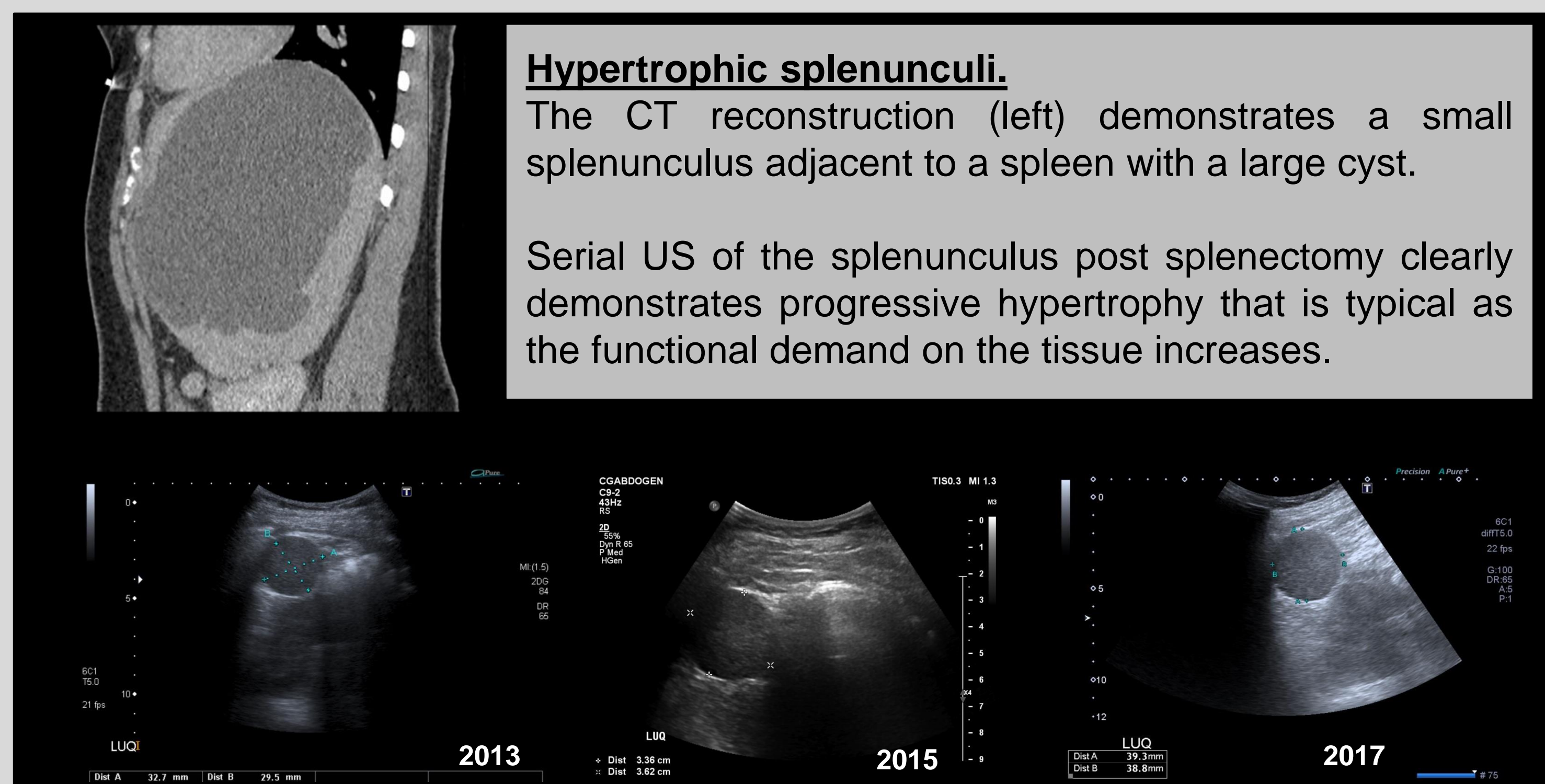
CEUS has recently been demonstrated to be of high value in the diagnosis due to the characteristic enhancement pattern of splenic tissue and has been endorsed by the European Federation of Societies for Ultrasound in Medicine and Biology (EFSUMB) in the Guidelines and Recommendations on the Clinical Practice of CEUS[1].

After injection, microbubbles remain entirely intravascular and are sequestered in both the liver and spleen[2]. When investigating suspected ectopic splenic tissue we are looking for persistent late phase enhancement (of 5 minutes or greater) as seen in the described cases.

Hypertrophic splenunculi.

The CT reconstruction (left) demonstrates a small splenunculus adjacent to a spleen with a large cyst.

Serial US of the splenunculus post splenectomy clearly demonstrates progressive hypertrophy that is typical as the functional demand on the tissue increases.

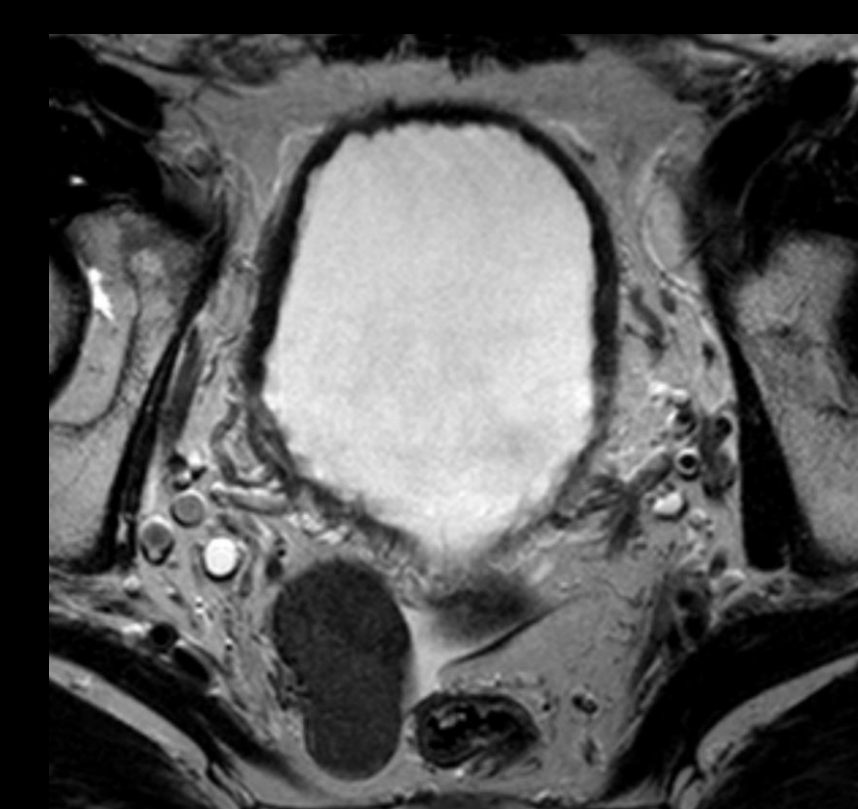


Pelvic splenosis

This pelvic splenosis deposit was found incidentally on a prostate MR. CT showed an irregular spleen which led to the suspicion of splenic trauma and splenosis.

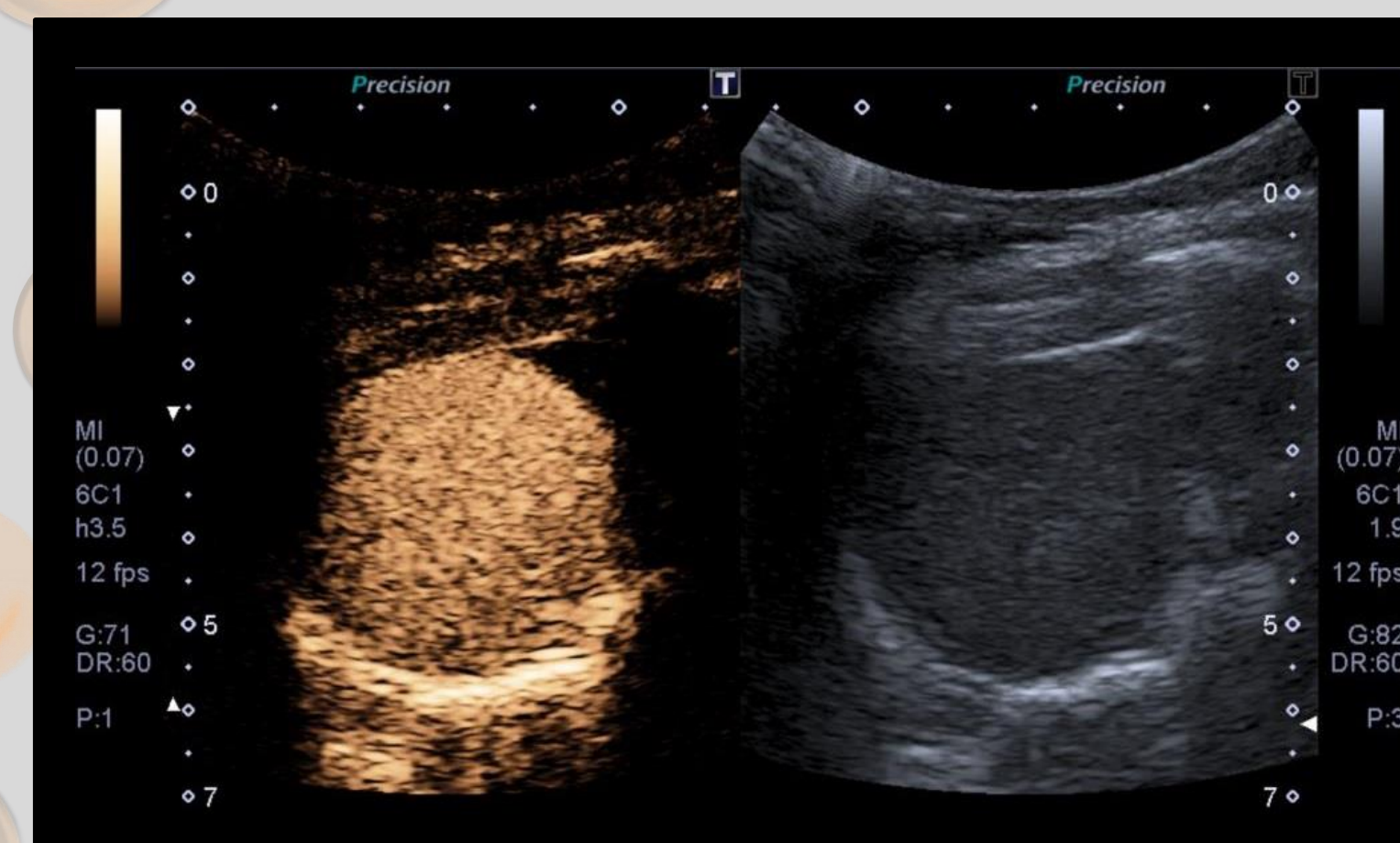
Diagnosis was confirmed by CEUS using the bladder as an acoustic window. There is persistent avid late phase enhancement (>6min post injection).

One of the key benefits of US is that it enables the practitioner to question the patient directly, something that can be invaluable in the absence of a detailed history.



Abdominal wall splenosis

CEUS showing enhancement of an abdominal wall splenosis following injection of ultrasound contrast microbubbles (SonoVue 2.4 mls). Both intense arterial enhancement and persistent late phase enhancement were observed and it remained strongly enhanced 5 minutes post injection.



References

- [1] Sidhu PS, Cantisani V, Dietrich CF, Gilja OH, Saftoiu A et al. The EFSUMB Guidelines and Recommendations for the Clinical Practice of Contrast-Enhanced Ultrasound (CEUS) in Non-Hepatic Applications: Update 2017. *Ultraschall in Med* 2018;39:2-44
- [2] Lim AKP, Patel N, Eckersley RJ, Taylor-Robinson SD, Cosgrove DO, Blomley MK. Evidence for spleen-specific uptake of a microbubble contrast agent: a quantitative study in healthy volunteers. *Radiology* 2004;231(3):785-788