Clinical History:

Asymptomatic elderly woman with unremarkable medical history including chronic autoimmune thyroiditis. Recent incidental finding of a sizeable, mixed hypoechoic splenic lesion at routine abdominal ultrasound, not reported in previous ultrasound studies performed years earlier. Normal findings at physical examination. No significant laboratory abnormalities (haemoglobin 12.8 g/dL; 210.000 platelets/mmc; normal coagulation).

Imaging Findings:

CT (Fig. 1) confirmed well-demarcated 6x4.5 cm ovoid mass at the upper splenic pole, mildly hypoattenuating compared to the splenic parenchyma, with inhomogeneous peripheral enhancement which progressed from the arterial over the portal venous and equilibrium phases. No other abnormal findings were present in the thorax, abdomen and pelvis.

Since malignancy could not be excluded, further investigation was requested. Six weeks later, MRI (Figs. 2, 3) confirmed solid splenic lesion with unchanged size, low peripheral T2-signal intensity, unrestricted diffusion with indeterminate apparent diffusion coefficient (ADC) value (0.9) and lesion-to-spleen ADC ratio (0.85). The dynamic post-gadolinium study confirmed inhomogeneous arterial hypervascularisation with progressive enhancement during the portal and venous phases, and central hyperenhancement in the equilibrium phase suggesting desmoplastic / fibrotic component.

Laparoscopic splenectomy was performed. The brownish-grey mass at cut section showed histopathology findings including myofibroblasts and associated heterogeneous inflammatory infiltrate with lymphocytes and plasma cells, consistent with inflammatory pseudotumour.

Discussion:

Inflammatory pseudotumours (IPTs) are uncommon benign tumour-like lesions which affect the orbit, lungs, and occasionally a variety of other organs including the gastrointestinal tract, liver, heart, bladder and thyroid. Splenic IPTs are exceedingly rare, with approximately 115 cases reported in recent literature review. The pathogenesis is debated, and possible aetiologies including immunological derangement, viral infections and parenchymal infarction have been speculated. Causative microorganisms have never been detected, albeit approximately 41% of cases tested positive for Epstein-Barr virus RNA [1-7].

Histologically, splenic IPTs are well-circumscribed solitary masses composed of myofibroblastic spindle cells,
intermixed with heterogeneous cellular infiltrate predominantly composed of plasma cells and lymphocytes. The key differential diagnosis is the exceptional follicular dendritic cell sarcoma, which is closely similar at imaging but shows atypical myofibroblasts and behaves more aggressively [1, 2, 6-8]. Splenic IPTs occur from 20 to 80 years of age in both sexes, is commonly asymptomatic and incidentally detected in two-thirds of cases because of increased use of imaging. Manifestations include nonspecific upper abdominal discomfort or pain, weight loss, sometimes fever and splenomegaly. Laboratory studies are normal or may indicate chronic inflammatory response with anaemia, thrombocytosis, polyclonal hypergammaglobulinaemia [1-7]. Splenic tumours are often challenging for the radiologist, and imaging does not allow confident differentiation of IPT from other lesions, particularly lymphoma and metastases. IPTs range from 3 to 22 cm, and commonly measure over 10 cm. Sonographically, IPTs appear as demarcated hypoechoic or partially calcified echogenic masses, hypovascular on colour-flow Doppler ultrasound. The CT appearance includes low- or isoattenuating lesions with variable, mild or heterogeneous early enhancement, gradual filling during the venous and delayed phases. The same pattern is demonstrated at dynamic contrast-enhanced MRI. IPTs have isointense T1- and either increased or decreased T2-weighted signal: as in this case, MRI may allow differentiation from strongly T2-hyperintense splenic masses such as haemangioma, lymphangioma, abscesses and necrotic tumours [3, 9]. On positron-emission tomography (PET)-CT these lesions show variable, often intense uptake [2]. Unfortunately IPTs generally masquerade as a splenic malignancy. Fine-needle and core biopsies are not recommended due to poor diagnostic yield, potential bleeding and fear of neoplastic dissemination. Although surgeons may be reluctant to remove asymptomatic masses, surgery is indicated even if IPT is suspected, since only histopathology establishes the diagnosis. Splenectomy is the mainstay treatment and increasingly performed laparoscopically. After splenectomy, the prognosis is favourable: metastases, local invasion or recurrence have never been reported [1, 3, 10-12].

**Differential Diagnosis List:** Inflammatory (myofibroblastic) pseudotumour of the spleen, Splenic haemangioma / hamartoma, Eosinophilic granuloma of the liver, Splenic abscess, Parasitic infestation, Lymphoma, Castleman’s disease, Metastasis from unknown primary tumour, Angiosarcoma, Follicular dendritic cell sarcoma lymphoma

**Final Diagnosis:** Inflammatory (myofibroblastic) pseudotumour of the spleen

**References:**


**Figure 1**

**a**

*Description:* Unenhanced acquisition confirmed ovoid mass (demarcated by arrowheads) at the upper splenic pole, mildly hypoattenuating compared to the remaining splenic parenchyma (*). *Origin:* Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)

**b**

*Description:* Arterial-phase (b,c) images showed inhomogeneous, predominantly peripheral enhancement of the demarcated splenic mass (arrowheads). *Origin:* Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** Arterial-phase (b,c) images showed inhomogeneous, predominantly peripheral enhancement of the demarcated splenic mass (arrowheads). **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Peripheral contrast enhancement progressed during the portal venous (d,e) phase. The ovoid splenic mass (arrowheads) appeared to be well-demarcated and measured approximately 4.5x6 cm. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Peripheral contrast enhancement progressed during the portal venous (d,e) phase. The ovoid splenic mass (arrowheads) appeared to be well-demarcated and measured approximately 4.5x6 cm. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Progressive persistent enhancement was seen in the delayed/equilibrium phase (f,g), where the upper pole splenic mass (arrowheads) appeared mostly isoattenuating compared to the splenic parenchyma (*) with minimal central inhomogeneity. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Progressive persistent enhancement was seen in the delayed/equilibrium phase (f,g), where the upper pole splenic mass (arrowheads) appeared mostly isoattenuating compared to the splenic parenchyma (*) with minimal central inhomogeneity. Origin: Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Coronal (a), axial (b) and fat-suppressed (c) T2-weighted images confirmed well-demarcated ovoid lesion (arrowheads) at the upper splenic pole, with lower signal intensity peripherally compared to the spleen (*). Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)

Description: T2-weighted images confirmed well-demarcated ovoid lesion (arrowheads) at the upper splenic pole, with lower signal intensity peripherally compared to the spleen (*) and some central inhomogeneity. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: T2-weighted images confirmed well-demarcated ovoid lesion (arrowheads) at the upper splenic pole, with lower signal intensity peripherally compared to the spleen (*) and some central inhomogeneity. **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)

Description: High (800) b-value diffusion-weighted acquisition showed unrestricted diffusion in the upper pole splenic mass (arrowheads). **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Corresponding apparent diffusion coefficient (ADC) map image showed hypointense splenic mass with median 0.9 ADC value. Lesion-to-spleen ADC ratio was approximately 0.85. **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)

Description: Corresponding axial T1-weighted image showed the mass lesion (arrowheads) to have similar low signal intensity compared to the splenic parenchyma. **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Figure 3

Description: Dynamic study including precontrast (a), late arterial (b), portal (c), venous (d,e) and equilibrium (f) phases was performed to further assess the upper splenic pole mass lesion (arrowheads). **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)

Description: The demarcated ovoid mass lesion (arrowheads) at the upper splenic pole was inhomogeneously vascularised during the arterial phase. **Origin:** Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Progressive enhancement of the splenic mass (arrowheads) was seen during the portal (c) and venous (d,e) phases. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Progressive enhancement of the splenic mass (arrowheads) was seen during the portal (c) and venous (d,e) phases. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)

Description: In the equilibrium phase the splenic mass lesion (arrowheads) showed persistent enhancement, centrally increased suggesting desmoplastic / fibrotic tissue. Origin: Tonolini M, Radiology Department, “Luigi Sacco” University Hospital – Milan (Italy)