CASE OF THE MONTH

An unusual perisplenic mass

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A female patient in her 70s was referred for image-guided biopsy of a perisplenic mass with malignant appearances.

She had presented six years previously with persistent ulcerating skin lesions on her lower legs, and dermatological histologists gave a differential diagnosis of cutaneous T-cell lymphoma or lymphomatoid papulosis. Lymphoma staging CT revealed splenomegaly and a 6×4 cm, well circumscribed, heterogeneously enhancing perisplenic mass. However, bone marrow biopsy was unremarkable and, after discussion with the patient,

haematologists elected to observe with serial CT imaging as they were unsure of the diagnosis.

Over the next five years the patient remained well and on CT the mass was seen to be gradually enlarging, but with no invasion or infiltration of adjacent structures.

However, six months prior to this referral, the patient developed a symptomatic pleural effusion, pulmonary hypertension on echocardiography, persistently elevated C-reactive protein, mild anaemia and considerable weight loss.

Figure 1 shows the CT findings at this stage and Figure 2 is representative of the ultrasound images.

(a)

(b)

Figure 1. (a, b) Contrast enhanced CT. An 11.4×8.2 cm heterogeneously enhancing mass is seen adjacent to the tip of the spleen. There are central areas of cystic degeneration/necrosis, ascites and splenomegaly, but no significant lymphadenopathy.

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Figure 2. Ultrasound examination with colour flow Doppler. The mass is hypoechoic, vascular and well defined. There is moderate ascites.

What is the differential diagnosis, and how should we proceed?

The differential diagnosis on the basis of clinical and radiological findings included lymphoma, hamartoma, gastrointestinal stromal tumour, vascular neoplasm, inflammatory pseudotumour and low-grade myofibroblastic sarcoma. Multidisciplinary team discussion agreed that a tissue diagnosis was required, but the patient was not keen for an open procedure, so percutaneous biopsy was performed under ultrasound guidance, without complication.

Histology showed no evidence of malignancy. It comprised bland spindle cells, many vessels and small primary lymphoid follicles, representing myofibroblastic proliferation without normal splenic sinusoids. These findings are consistent with an inflammatory pseudotumour.

Inflammatory pseudotumour

Inflammatory pseudotumours, also known as inflammatory myofibroblastic tumours, are generally considered to be benign, although the World Health Organization classification puts them in an intermediate category between benign and malignant ("rarely metastasising, <5%") [1]. They are usually solitary and have been reported in many different sites throughout the body, most commonly in the lung and orbit. In 1999, a literature search by Moriyami et al [2] revealed 67 reports of occurrence in the spleen, but we have been unable to find any reports of perisplenic inflammatory pseudotumours. Perhaps this patient's perisplenic pseudotumour arose from a splenunculus.

The clinical and radiological presentations of inflammatory pseudotumours often simulate a malignant process and diagnosis is often made on excision. It is frequently associated with splenomegaly, leukocytosis, anaemia or hypergammaglobulinaemia. There is no known association with pulmonary hypertension, as was found in this case, and the cause for this is not clear. Oshiro et al [1], however, reported a splenic pseudotumour associated with sleep apnoea syndrome and chronic bronchitis. They postulated that overexpression of certain cytokines demonstrated in inflammatory pseudotumours had contributed to the inflammatory lung disorder and sleep-related breathing disorder. After excision of the pseudotumour the sleep apnoea and bronchitis resolved. An undiagnosed inflammatory respiratory disorder or sleep disorder could account for the pulmonary hypertension in our patient. The literature would suggest that the ascites and pleural effusions are likely to be exudative secondary to the inflammation, and this was confirmed on diagnostic aspiration.

The aetiology of pseudotumours remains uncertain, although there is some evidence of the involvement of the Epstein-Barr virus in cases affecting the spleen, and of overproduction of interleukin-1 [3, 4]. Splenic pseudotumours have been reported in association with idiopathic thrombocytopenic purpura and have some architectural similarities with Riedel's thyroiditis and idiopathic retroperitoneal fibrosis, suggesting an autoimmune pathogenesis [5].

The imaging findings are variable and non-specific. On ultrasound, pseudotumours tend to be hypoechoic. Calcification may be present and a central scar has been described in several case reports. CT contrast enhancement and MR signal characteristics are variable, making pre-operative diagnosis difficult. The role of positron emission tomography scanning has yet to be established.

Percutaneous-guided biopsy is often inconclusive, but tissue diagnosis is usually required because of the non-specific clinical, radiological and laboratory data. In our case, the patient was not keen on an open procedure so biopsy was performed and the diagnosis made.

Conclusion

This case represents the first perisplenic pseudotumour described. Splenic pseudotumours are a difficult diagnosis to make, but many case reports exist. They are normally diagnosed post operatively because of the non-specific clinical and radiological findings. This case is also unusual, therefore, in that diagnosis was made without an open procedure. The six-year follow-up, without excision, after the lesion was first seen on CT also gives some insight into the natural history of this rare pathology. The presence of pulmonary hypertension in this case of inflammatory pseudotumour is also previously undescribed, and a possible biological mechanism has been conjectured.

There remains the question of treatment. The patient is currently feeling some improvement on a trial of steroids and there are multiple reports in the literature of cure after splenectomy in cases of splenic pseudotumours [6]. Should we advise removal of the mass in this case given the comorbidities, vascular nature of the pseudotumour and the patient's reluctance? Embolisation of pseudotumours is undescribed, but should it be considered?

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