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Epstein–Barr virus positive inflammatory pseudo-tumour of the spleen: A case report and literature review☆

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ABSTRACT

INTRODUCTION: Epstein–Barr virus positive inflammatory pseudo-tumour (IPT) of the spleen is an uncommon, frequently asymptomatic entity, which is typically picked up as an incidental finding on imaging.
PRESENTATION OF CASE: We present a case of EBV positive IPT of the spleen which presented as an incidental finding on CT in a patient with a history of malignancy. Splenectomy was performed.
DISCUSSION: IPTs are benign spindle cell lesions of varying aetiology, which can arise in a variety of tissues, including the spleen. In situ hybridisation showed strong staining for Epstein–Barr virus RNA in our case, in common with many similar lesions described in the literature. The differential diagnosis of such spindle cell tumours is discussed.
CONCLUSION: Radiologically, EBV positive spindle cell tumours are indistinguishable from malignant lesions such as lymphoma and diagnosis is made on histology, usually at splenectomy.

1. Introduction

Splenic tumours are an uncommon entity and include a wide range of both benign and malignant conditions. Haemangiomas are the most common benign splenic tumours, while lymphoma is the most frequently encountered malignancy. Less common conditions include littoral cell angioma, hamartoma and metastases.[1]

While ultrasound, CT and MRI are useful modalities for detecting splenic tumours, definitive diagnosis can usually only be made following splenectomy.

In this case report we present the clinical and pathological features of an inflammatory pseudotumour of the spleen, associated with Epstein–Barr virus and review the current classification of such splenic spindle cell lesions.

2. Case report

A 69-year-old lady with a history of breast and uterine carcinoma was found to have a mass at the splenic hilum on surveillance CT. In 2003 she had a hysterectomy for a uterine carcinoma but required no adjuvant treatment. In 2011 she presented with a right sided malignant breast lump and underwent a wide local excision and sentinel node biopsy. The lesion was a T2N0 Grade 2 breast carcinoma. She was commenced on Arimidex post-operatively. A routine follow-up CT scan of the abdomen demonstrated a 7.2 cm × 7.4 cm mixed attenuation, solid rim-enhancing mass lesion in the hilum of the spleen (Figs. 1 and 2). At the time of her CT scan she was virtually asymptomatic with the exception of some 'fullness in the upper abdomen'. Her case was discussed at the oeophago-gastric multi-disciplinary team meeting and a differential diagnosis of lymphoma or metastatic deposit was given. Percutaneous biopsy was not felt appropriate given the risk of bleeding although FNA and needle core of the spleen has been described.[2]

She underwent an uneventful splenectomy in October 2011. On sectioning the normal sized spleen there was a rubbery, well-defined tumour measuring 8 cm × 6.5 cm × 5 cm (Fig. 3). Histology showed a tumour composed of spindle cells, intimately admixed with lymphocytes, plasma cells and occasional eosinophils (Fig. 4). In the centre of the lesion there were areas of coagulative tumour necrosis but minimal evidence of nuclear atypia or anaplasia. In situ hybridisation for Epstein–Barr virus revealed Epstein–Barr virus encoded RNA (EBER) in all of the spindle tumour cells (Fig. 5). Immunohistochemical stains for vimentin and smooth muscle actin were strongly and widely positive. The follicular dendritic cell marker CD21 was negative but CD23, CD35 and fascin were focally positive.

She had an uncomplicated post-operative recovery and at her 6-month follow-up appointment she was well and symptom free.

3. Discussion

"Inflammatory pseudotumor" (IPT) is a term that has been used to describe a variety of tumefactive lesions of varying aetiology and different behaviour in a variety of anatomical sites including inflammatory myofibroblastic tumour (IMT; a low grade

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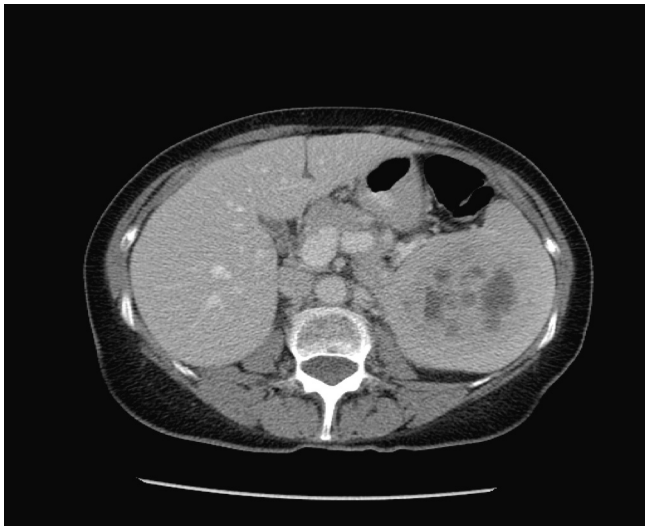

Fig. 1. Axial CT image showing splenic mass.

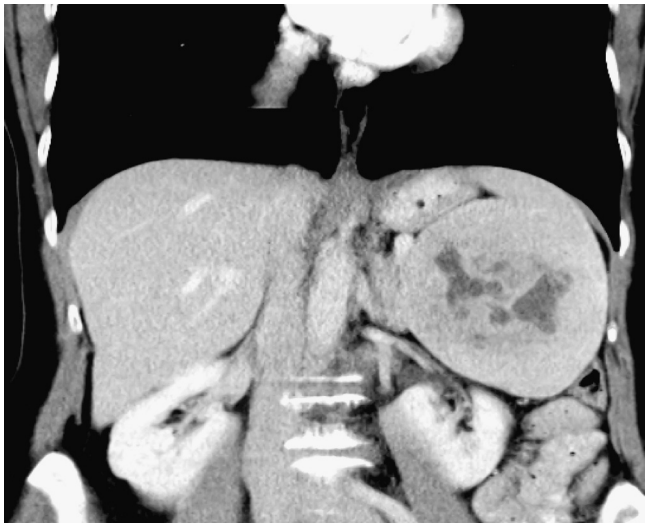

Fig. 2. Coronal CT image of splenic mass.

Fig. 3. A section through the spleen shows a well circumscribed, firm, white tumour mass measuring 8 cm in diameter.

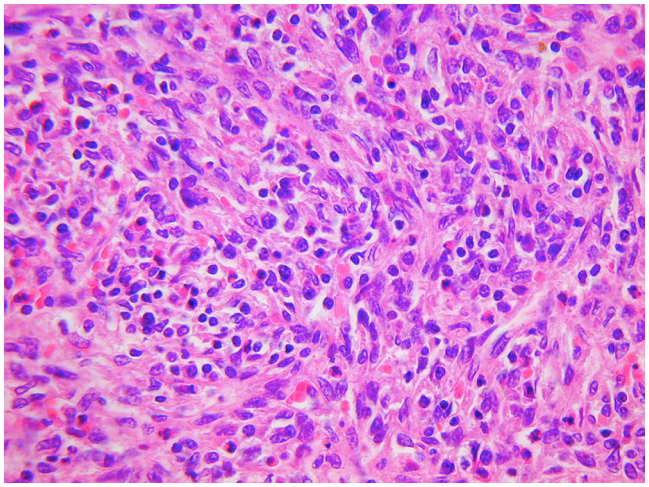

Fig. 4. Haematoxylin and eosin stain (original magnification 200×) showing rather spindled cells a background of lymphocytes, plasma cells and scattered eosinophils.

malignancy commoner in younger patients and associated with abnormal ALK-1 expression) infective mycobacterial spindle cell proliferations and the tumefactive complications of IgG4 related disease. Follicular dendritic cell tumour is a rare, low grade malignancy that most commonly occurs as a primary lymph node tumour.[3] However, it has been reported in a variety of extra nodal areas such as the oral cavity, lung, tonsil, pharynx and thyroid where it is usually EBV negative.

A specific association between hepatic and splenic IPTs and Epstein–Barr virus has been proposed, with 40–67% of cases testing positive for the virus.[4,5] In a study of 18 cases Arber et al.,[4] identified EBV nucleic acid in the spindle cell population, within the IPT. The spindle cells were uniformly vimentin positive and in some cases they were smooth muscle actin positive (SMA), suggestive of myofibroblastic differentiation.

In 2001 Neuhauser examined 12 splenic IPTs and found that 50% were positive for EBER. SMA was the most commonly expressed marker followed by vimentin.[6] Notably 3 of the 10 cases were associated with a past medical history of carcinoma and 2 of the 10 cases were positive for the follicular dendritic cell marker CD21.

In 2001, Cheuk reported 11 splenic tumours where the spindle cells showed positive staining for EBER and the follicular dendritic cell markers CD21/CD35 and CD23 and proposed the term

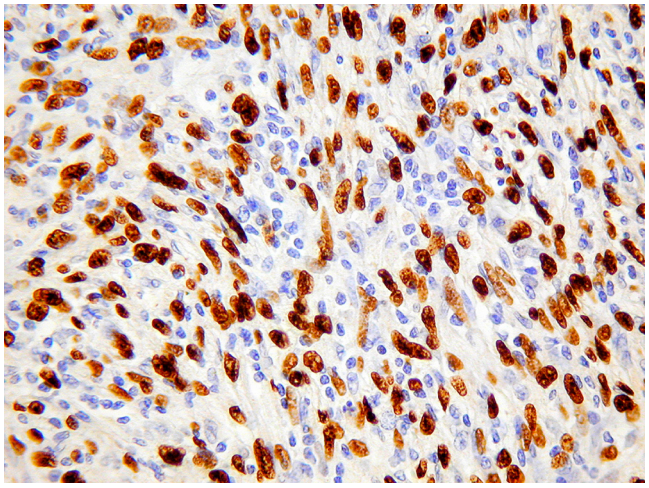

Fig. 5. EBER in situ hybridisation showing Epstein–Barr virus encoded RNA, predominantly in the spindle cells, with no staining in the smaller lymphocytes (original magnification 400×).

"EBV related inflammatory pseudotumour like follicular dendritic cell tumour of the spleen" which were characterised by indolent behaviour (although there were some recurrences) and a female predeliction.[7]

1n 2003, Krishnan and Frizzera[8] proposed categorising splenic spindle cell tumours into three groups:

(1) IPT like FDC tumour – histology of dispersed FDCs in a prominent inflammatory background; and consistent evidence of EBV infection of the spindle cells.
(2) IMT of spleen – similar to IMTS in other sites. A good proportion (66%) is EBV positive. Follow up for splenic IMTs uneventful compared to other locations.
(3) IPT of spleen – predominant spindle cells are not SMA positive MFs. No evidence of EBV infection.

"Splenic stromal tumour" has been proposed as an alternative term for this heterogenous group of tumours[9] and indeed other authors have observed that such lesions may show variable follicular dendritic or myofibroblastic differentiation and that this may be associated with the levels of EBV expression.[10]

4. Conclusion

EBV positive inflammatory pseudotumour of the spleen is an uncommon spindle cell tumour of the spleen which can show variable myofibroblastic or follicular dendritic differentiation and variation in expression levels of EBV These lesions may be associated with prior malignancy and in the era of increased surveillance with cross sectional imaging may present in asymptomatic patients with radiological findings suggestive of lymphoma or metastatic disease. Diagnosis is usually made at splenectomy.

Conflict of interest statement

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for the publication of this case report and images and a copy is available for review.

Author contributions

Dr Paula Loughlin, Dr Eimear Devlin, Dr Aidan Brady, Dr Damian McManus and Professor Roy Spence all contributed to the writing, editing and literature review. Dr Eoin Napier contributed the radiology images.

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