Littoral Cell Angioma of the Spleen: a case report

Gabriella Grech, Angela Sultana

Abstract

The patient is a 54 year old, presenting with a 4 month history of worsening lower back pain, radiating to both lower limbs. CT Thorax Abdomen and Pelvis was carried out which showed a 6.2cm dense fluid density lesion in the spleen. The case was discussed at the multidisciplinary team meeting and open splenectomy was carried out. Histological diagnosis was consistent with an infarcted littoral cell angioma.

Key words:

Littoral cell angioma, benign, spleen, splenectomy, histology

Littoral cell angioma is a rare vascular tumour unique to the spleen, originating from cells lining the venous sinuses of the normal spleen. It was first described by Falk et al. in 1991. The majority of cases have been composed of multiple nodules of varying sizes in the spleen, benign and asymptomatic in nature.

Case Report:

The patient is a 54 year old female, presenting with a 4 month history of worsening lower back pain, radiating to both lower limbs. The pain was associated with paraesthesia, weight loss and loss of appetite. She was noted to have a high white cell count and C-reactive protein.

A Computerised Tomography (CT) Scan of the Thorax, Abdomen and Pelvis was carried out which showed a 6.2cm dense fluid density lesion in the spleen. This demonstrated peripheral enhancement with internal septations. There was no intraperitoneal rupture. The splenic capsule was intact and there was no subcapsular extension. No further splenic lesions noted. The main differential diagnoses were splenic abscess, hydatid cyst and neoplastic tumour.

The CT Scan also showed bilateral sternoclavicular joint erosion with osteitis, multiple endplate sclerotic foci and bilateral sacroilitis. These findings explained the symptoms that the patient presented with. She was diagnosed with SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis and osteitis) and referred to rheumatology.

Ultrasoundography of the splenic lesion revealed a heterogenous lesion with a well-defined wall, internal septations and cystic areas, in keeping with an abscess. The patient was started on intravenous antibiotics and an ultrasound-guided drainage of the abscess was carried out. A sample of pus was sent...
for microscopy, culture and sensitivity. Microscopy showed few polymorphs and no bacteria were seen or cultivated. The case was discussed at the multidisciplinary team meeting and open splenectomy was planned for the patient for definite diagnosis of the splenic lesion.

Histology showed a spleen measuring 100mm X 78mm x 75mm. On sectioning there was a well circumscribed lesion measuring 50mm in maximum dimension. The lesion was partly cystic and partly solid. The cystic component contained garish, yellow viscous material and solid component had a pale yellow cut surface.

On microscopic examination, the lesion consisted of abundant necrosis, acute and chronic inflammatory cells. Admixed amongst the necrotic debris and most prominent at the interface between viable splenic tissue and the necrotic area were plump vaculated histiocyte like cells, containing eosinophilic material. The background spleen appeared unremarkable. The cells were highlighted by CD68, CD31 and are negative for CD34 (different pattern of staining which highlights vessels). Stains for MNF116, CD1a were negative. S100 probably highlighted endogenous activated macrophages. Special stains for iron, fungal organisms and acid fast bacilli were negative. Diagnosis was consistent with an infarcted littoral cell angioma.

Literature review:

This tumour occurs mostly in middle-aged men and women and has equal sex distribution. Several studies have shown associations of littoral cell angioma of the spleen with immunological or congenital disorders such as Crohn’s disease, Wiskott-Aldrich syndrome, Epstein syndrome, lymphocytic colitis, ankylosing spondylitis, Gaucher’s disease, myelodysplastic syndrome, chronic glomerulonephritis or aplastic anaemia. Moreover, one-third of cases are associated with tumours of visceral organs such as colorectal, renal, hepatocellular, lung and pancreatic adenocarcinoma and therefore close clinical follow-up of these patients is recommended.

Differential diagnosis of multinodular splenomegaly includes multiple haemangiomases, lymphangioma, hamartoma, haemangiopericytoma, hemangiendothelioma, angiosarcoma, lymphoma, metastatic disease, Kaposi’s sarcoma and disseminated infections caused by fungi, mycobacteria, pneumocystis carinii and sarcoidosis.

Radiological findings are rarely sufficient for making a definite diagnosis of littoral cell angioma of the spleen. On ultrasound examination, the appearance of these tumours is variable. It includes mottled ecotexture without discrete lesions as well as isoechoic, hypoechoic and hyperechoic lesions. On abdominal CT Scans, littoral cell angioma typically manifests as multiple hypoattenuating lesions that enhance homogeneously or inhomogeneously as these are vascular tumours. On Magnetic Resonance Imaging (MRI), the nodular lesions of littoral cell angioma typically appears markedly hypointense with both T1 and T2 weighted pulse sequences. This reflects the presence of haemosiderin in the lesions due to the haemophagocytic capacity of the neoplastic cells. The gold standard treatment is splenectomy. Reports of other treatments including glucocorticoids and angioembolisation have been published. The definitive diagnosis is made on histology and confirmed with immunohistochemistry. There is a mixture of papillary and cystic areas. Neoplastic cells derived from normal splenic lining-littoral cells form the lining of these papillary and cystic areas. This neoplasm has features of both endothelial and histiocytic differentiation with the typical and characteristic immunohistochemical pattern of littoral cell angioma being CD31, CD68, CD163, CD21, FVIII antigen positive; CD34, CD8 negative.

Littoral cell angioma is a benign tumour of the spleen, which may be associated with malignancy, immunological and congenital disorders. The treatment of choice is splenectomy. The imaging features of many other splenic neoplasms may mimic those of littoral cell angioma but in such cases diagnostic signs and symptoms are usually present. In cases of incidental finding of splenic mass on imaging and the patient has no associated signs or symptoms, littoral cell angioma should be suspected.
References:


