Splenic Littoral cell angioma in a patient with myasthenia gravis

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ABSTRACT

Littoral cell angioma (LCA) is a rare primary vascular tumour of the spleen. LCA has been associated with autoimmune disorders and it is hypothesised that the underlying pathogenesis is immune dysregulation. We report a case of LCA in a patient with Myasthenia Gravis which has thus far not been reported in the literature.

Key Words: Littoral cell angioma, myasthenia gravis, spleen, neoplasm

INTRODUCTION

Littoral cell angioma (LCA) is a rare primary tumour of the spleen that has been first described in 1991. 1 LCA has unique histological and immunophenotypic features and has been associated with malignancy and autoimmune disorders. Definitive diagnosis is usually made on obtaining splenectomy specimens postoperatively. We report a case of LCA in an elderly lady who recently has been diagnosed with Myasthenia Gravis. The diagnosis of LCA was made on percutaneous biopsy of the lesion. The association of LCA and Myasthenia Gravis has thus far not been reported in the literature.

CASE REPORT

A 77-year-old female patient with a history of euthyroid multinodular goitre, presented with bilateral ptosis and bulbar palsy. Following a series of investigations, she was diagnosed to have Myasthenia Gravis (MG). She subsequently underwent a Computed Tomography (CT) scan of her neck and thorax to look for evidence of thymoma. The CT scan incidentally revealed an enlarged spleen with a solitary lesion measuring 7.1 x 6.0 x 7.8 cm (width x anterior posterior x craniocaudal). There was no definite capsule surrounding the lesion, and it contained multiple hypoattenuating areas of varying sizes ranging between three and 12 mm with no calcific foci within. The enlarged spleen displaced the splenic vein anteriorly with splaying of the splenic hilum. A clear fat plane could be seen between the lesion and the splenic vessels anteriorly, left...
adrenal gland medially, left kidney postero-inferiorly and the tail of the pancreas (Figure 1). There was no associated lymphadenopathy or liver lesions seen. On further questioning the patient denied any abdominal or constitutional symptoms. Her blood parameters including her platelets count was within normal limits.

The splenic mass was subsequently biopsied percutaneously under ultrasound guidance and histological examination revealed that the lesion was composed of anastomosing vascular channels which were lined by endothelial cells (Figure 2a). These cells expressed both histiocytic (CD68) and endothelial (Factor 8) markers (Figure 2b and c). Therefore a diagnosis of littoral cell angioma was made. After discussion and in view of her age and being without symptoms, the patient opted for conservative management of the splenic mass.

A magnetic resonance imaging (MRI) was six years following the initial CT Scan showed that the mass had increased in size measuring 14.9 x 12.0 x 16.8 cm (Figure 3). Multiple ovoid areas seen within the lesion were hypointense on T1 weighted sequence and mostly hyperintense on T2 weighted sequence. Some of the areas which were hypointense in T2 weighted sequence likely represented hemosiderin deposition within. As seen before, there was neither involvement of the adjacent organs or the splenic vessels but the surrounding organs were displaced secondary to the mass effect (Figure 3). The patient was also relatively asymptomatic apart from having a distended abdomen with a palpable mass at the left upper quadrant.

**DISCUSSION**

LCA is a rare primary vascular tumour of the spleen and was first described by Falk et al. in 1991. ¹ LCA originates from the specialised

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¹ Falk et al., 1991.
endothelial cells lining the sinus channels of the splenic red pulp, called "littoral cells", which are unique among vascular lining cells in that they express histiocytic antigens and the usual endothelial antigens. LCA presents in two forms, the more common multiple nodular form as in our patient and the rarer solitary form.

There is no age and gender predilection, however in the original paper by Falk et al. the median age at diagnosis was 49 years. The majority of patients with LCA are usually asymptomatic and the lesion is discovered incidentally as was the case with our patient. However, some patients may present with abdominal pain, pyrexia of unknown origin, weight loss, splenomegaly and blood investigation may reveal thrombocytopenia and anaemia secondary to hypersplenism.

The pathogenesis of LCA remains unclear, but a hypothesis of immune system dysregulation has been postulated given its association with autoimmune disorders such as Crohn's disease and Gaucher's Disease. In our case, our patient had MG that was just previously diagnosed before the finding of LCA. Ours is the first reported case of LCA in association with MG in the literature. A strong oncogenic association has also been reported with LCA. Associated malignancies reported are colorectal adenocarcinoma, pancreatic cystadenocarcinoma, renal cell carcinoma, non-small cell cancer of lung, hepatocellular carcinoma and transitional cell carcinoma of the bladder. Although LCA was initially thought to be a benign neoplasm, two malignant subtypes have been described which include littoral cell angiosarcoma and littoral cell hemagioendothelioma. Due to the association with other organ malignancies coupled with LCA exhibiting malignant potential, close monitoring and surveillance of patients have been recommended.

Several radiographic tools can be utilised in evaluating LCA, although a definitive diagnosis essentially requires histological confirmation. Ultrasonography features of LCA is diversified, and findings of isoechoic, hypoechoic and hyperechoic lesions have been reported. CT features of LCA include the presence of multiple splenic masses which are low attenuating on early contrast enhanced images, and isodensitometric lesions on delayed contrast images. Delayed contrast material filling of LCA compared to normal splenic parenchyma suggests that the abnormal vascular channels that form LCA become perfused with contrast at a slower rate than the normal splenic parenchyma. On MRI, the splenic mass (es) typically appear hyperintense in T2-weighted images as with our patient although in a minority of cases both T1
and T2 weighted images show hypointense lesions due to the haemosiderin content. 4

A definitive diagnosis of LCA requires histological examination. Immunohistochemical staining of LCA exhibit positive staining for endothelial (factor VIII, CD31), histiocytic (CD68 and lysozyme) and complement receptors (CD 21 and CD 163). 2, 4 LCA is typically negative for CD8, CD34 and S-100 protein antigens. 1, 2 Our patient underwent a fine needle biopsy of the lesion to obtain a tissue diagnosis. This option for obtaining tissue is rarely reported in the literature as complications of bleeding is highly anticipated as spleen is a vascular organ. However, the reported complication rate with imaging guided biopsy of splenic lesions is between 0 - 2%. 11 A higher rate of 10.3% has been reported in patients with refractory thrombocytopenia. 12

Treatment for symptomatic LCA is splenectomy, but currently no recommendations for asymptomatic LCA. 2 Due to the association and reported cases of malignant transformation, the option for splenectomy should be considered. Our patient opted not to have splenectomy due to her age and comorbidities and after six years of follow up she remains relatively symptom free with no evidence of malignant transformation.

In conclusion, LCA is a rare vascular tumour of the spleen which is often found incidentally as patients are usually asymptomatic. However, LCA is associated with malignant conditions and several autoimmune disorders including MG which we have highlighted. It is feasible to make a diagnosis of LCA on percutaneous biopsy of the lesion with minimal risk.

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