Littoral Cell Angioma Of Spleen: A Case Report

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Abstract: Littoral cell angioma (LCA) is a rare primary vascular tumor of spleen first described in 1991 by Falk et al. It originates from the cells lining the venous sinuses of the normal spleen. These tumors are unique in that they display both epithelial and histiocytic properties based on their cell of origin, the splenic littoral cells. LCA is usually benign but cases of malignant nature has also been reported. There is a strong association between this neoplasm and a group of immunologic or oncologic entities, including Crohn’s disease and adenosarcoma of colon and pancreas.

The clinical representation of LCA ranges from being completely asymptomatic and discovered incidentally, to presenting with a constellation of signs and symptoms such as abdominal distension, complex constitutional symptoms, splenomegaly, and hypersplenism. Although the computed tomography (CT) and ultrasound (US) features of this neoplasm have been well described, there is a lack of specificity in differentiating the tumor from other primary vascular tumors of spleen. The definitive diagnosis can only be made after histological and immuno-histochemical studies. Splenectomy remains the treatment of choice.

I. Introduction

Littoral cell angioma (LCA) is a rare primary vascular tumor of spleen first described in 1991 by Falk et al. It originates from the cells lining the venous sinuses of the normal spleen. These tumors are unique in that they display both epithelial and histiocytic properties based on their cell of origin, the splenic littoral cells. LCA is usually benign but cases of malignant nature has also been reported. There is a strong association between this neoplasm and a group of immunologic or oncologic entities, including Crohn’s disease and adenosarcoma of colon and pancreas.

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II. Case report

A 65 year old male presented to us with complaints of fever, weight loss, generalized body weakness, fatigability and dragging pain in left upper abdomen since last 6 months. Patient was evaluated for the same and USG abdomen showed mild splenomegaly with multiple hypo-echoic lesions throughout the spleen. For further evaluation CT scan abdomen was done which showed multiple, round, hypo-dense lesions with suspicion of malignant etiology. To confirm the malignant nature, USG guided FNAC was done which came out to be inconclusive. Rest all routine blood investigations (CBC, PBF, biochemistry) were within normal limits. Considering the possibility of malignant nature of disease splenectomy was planned which was done through minimal invasive approach. Intra-operative findings were:- mild splenomegaly with multiple round lesion scattered all over the spleen. Post-operative recovery was uneventful and patient was discharged 2 days later. Histologically, lesion was described as a vascular neoplasm with anastomosing vascular channels lined by histiococytes with occasional papillary structures, which was consistent with LCA.

III. Discussion

Primary vascular tumors of the spleen are uncommon but represent the majority of non-hematolymphoid splenic tumors. The differential diagnosis of splenic vascular tumors is broad and may represent benign (haemangioma, haemartoma, lymphangioma), indeterminate (littoral cell angioma, haemangioidoethelioma, haemangiopericytoma), or malignant neoplasms (angiosarcoma). LCA is a recently described vascular tumor of spleen that is now classified as having uncertain biological behaviour, given several case reports which have identified malignant potential. It originates from the cells lining the venous sinuses of the normal spleen, which, under yet unexplained stimuli, proliferate to form the characteristic lesions seen grossly. It has been postulated that these cells react to as yet unknown antigenic stimuli with proliferation and increasing phagocytic activity as a result of their unusual elongated cytoplasmic surface and basement membrane discontinuity. The pathogenesis of LCA remains unclear, but given its association with autoimmune disorders such as Crohn's disease and inborn metabolic diseases such as Gaucher's disease, immune system dysfunction has been postulated as a possible important pathogenic mechanism. Supporting this hypothesis, other reports have suggested that chronic infection and systemic immuno-suppression may contribute to LCA development.

The exact incidence of LCA is unknown although the incidence of splenic haemangioma varies from 0.03% to as high as 14% in a series of autopsy reports. LCA does not have any particular gender or age predilection although the median age in Falk et al.'s original study of LCA was 49 years. LCA may be completely asymptomatic and represent an incidental finding by imaging. LCA may also present with a myriad of possible signs and symptoms, such as:

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spleenomegaly with or without abdominal pain, hypersplenism with ensuing anaemia and/or thrombocytopenia, and constitutional symptoms such as intermittent fevers. More dramatically, LCA has been reported to present as splenic rupture and haemoperitoneum. Indeed, immune system dysregulation may explain the association of LCA with other cancer types. The other cancer types associated with LCA include: thyroid, colorectal, renal, pancreatic, hematologic (lymphoma), ovarian, and testicular cancer. These observations have prompted recommendations to closely evaluate and provide surveillance to patients with LCA for the development of other malignancies. Conversely, the association of LCA with other cancer types may also be a result of making an incidental diagnosis of LCA during extensive radiological imaging for other diseases, given the largely asymptomatic presentation of these tumors. However, close follow-up of LCA may be warranted due to their potential for malignant transformation. The two subtypes of LCA with malignant potential have been described as "littoral cell angiosarcoma" and "littoral cell haemangioendothelioma". These LCA variants may present with distant metastasis several months after splenectomy; histologic evaluation reveals features consistent with LCA histopathology as well as abnormal architecture, nuclear atypia, and necrosis.

Radiologically, LCA may be evaluated by several imaging modalities such as US, CT, MRI, or nuclear medicine studies (Tc-99m labelled RBC scintigraphy). US may reveal lobular splenomegaly with heterogeneous nodules (either hypo- or hyper-echoic) that may be solitary or multiple. On non-contrast CT, LCA appear as hypo-attenuating masses; given the vascular nature of these neoplasms, they tend to enhance homogeneously. On MRI, a minority of cases may be hypointense on both T1-weighted and T2-weighted scans because of hemosiderin content of the tumor. However, as significant siderosis is seen in less than 50% of LCA cases, lesions tend to be hyperintense on the T2 weighted images. Nuclear medicine studies with Tc-99m labelled RBC scintigraphy can be useful to differentiate splenic lesions from splenic haemangiommas. However, the radiologic features of LCA are rarely diagnostic since many other splenic neoplasms such as haematomas, haemangiomas, lymphomas, metastatic disease and infectious processes exhibit similar imaging characteristics.

Grossly, there are 2 forms of LCA. More commonly it is seen as tumor consisting of multiple nodules diffusely involving the entire spleen, however a rare solitary form of LCA has also been described. The colour of these nodules may be dark red, brown, or black, consistent with blood or blood products of varying chronicity. Rarely, LCA appears white on gross pathology.

Microscopically, there are several distinguishing histological and molecular features of LCA. Histologically, LCA has specific features that differentiate it from other primary vascular tumors, including angiosarcoma. LCA are composed of anastomosing vascular channels resembling splenic sinusoids and have irregular lumina featuring papillary projections and cyst-like spaces. Tall endothelial cells with histiocytic properties that slough off into the vascular lumen are common, as the absence of atypical cells and presence of low mitotic activity. By immuno-histochemical staining, these tumor cells will express endothelial and histiocyte antigens, a reflection of the distinct dual differentiation potential of LCA. Such expression includes endothelial markers (factor VIII Ag and CD 31/BMA 120) as well as histiocytic markers (CD 68/KP I and lysozyme). The expression of these molecular markers has also been demonstrated in fine-needle aspiration biopsies of LCA.

Symptomatic LCA are often relieved by splenectomy, and given the association of LCA with other malignancies and reported cases of metastasizing LCA, splenectomy is both diagnostic and therapeutic. While there have been reports of medical therapy with gluco-corticoids and angio-embolization of splenic haemangiommas, splenectomy is still considered the gold standard for treatment of vascular splenic tumors.

IV. Conclusion

LCA is a recently described primary vascular neoplasm of the spleen that may be associated with other malignancies and may itself also have malignant potential. Several radiological studies may suggest LCA, although a pathological diagnosis, either by core biopsy or diagnostic splenectomy is imperative. This rare case illustrates the importance of thoroughly evaluating incidental vascular splenic tumors. Although the vast majority of LCAs are benign, their differential diagnosis must include both primary and secondary malignancy, given LCA’s association with other cancer types as well as their uncertain malignant potential. With this in mind, gold standard management remains splenectomy and long-term follow-up for the development of synchronous tumors or metastic lesions is advised.

References


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