Case Report

Littoral Cell Angioma: A Rare Cause of Pediatric Thrombocytopenia

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Abstract

Littoral cell angioma is a rare primary splenic vascular tumor, originating from the littoral cells lining the red pulp sinuses of the spleen, with variable presentation. Here, we present the case of a 2-year-old male child presenting with chronic thrombocytopenia which resolved after splenectomy.

Keywords: *Littoral cell angioma, splenectomy, thrombocytopenia*

Introduction

Littoral cell angioma (LCA) is a rare primary splenic vascular tumor that originates from the splenic littoral cells. The term "littoral" is derived from the Latin noun littoris, meaning shore. Littoral cells typically line the splenic red pulp sinuses. It has been postulated that these cells react to unknown antigenic stimuli with proliferation and increasing phagocytic activity as a result of their unusual elongated cytoplasmic surface and basement membrane discontinuity. Usually on gross pathology, a large spleen with multiple nodular lesions is common, whereas on histopathology LCA is based on the presence of anastomosing vascular channels of variable size lined with flat and tall endothelial cells, focal papillary fronds extended into the vascular channels and normal splenic sinuses at the periphery of the lesions in the splenic red pulp. It is mostly seen in the adult population. It is commonly associated with adenocarcinoma of the colon and pancreas and immunologic entities such as Crohn's disease.

Case Report

A 2-year-old male child was brought to us with complaints of recurrent episodes of epistaxis and persistent purpuric patches over various body parts since 8 months. On examination, he was hemodynamically stable with spleen of 2 cm below costal margin without any significant lymphadenopathy and bony tenderness. The child had no history of fever, weight loss or measles, mumps, and rubella vaccination. On reviewing his past blood reports, his platelet counts were persistently in range of 20,000–50,000/mL with normal leukocyte count and moderate grade anemia (Hb-10.6 g%). He had a negative antinuclear antibody test report. Bone marrow biopsy and peripheral smear showed giant platelets consistent with hypersplenism. The child was given platelet transfusion and was considered for splenectomy. While the child was awaiting splenectomy, he encountered an episode of acute abdominal pain. Radiological workup showed large splenic subcapsular hematoma measuring 5 cm × 4.5 cm. An emergency splenectomy was carried out. Histopathological study of expunged tissue showed LCA. He had an uneventful postoperative period, and after 6 years of follow-up, the child is healthy.

Discussion

LCA, first defined in 1991 by Falk *et al*.[1] is a rare, benign vascular tumor of the spleen. LCA originates from the littoral cells that line the sinusoids of the red pulp. LCA expressed endothelial and histiocyte associated antigens, similar to littoral cells lining venous sinuses of normal spleen. The incidence of splenic hemangioma varies from 0.03% to 14% at the autopsy series.[2] Although there is no age predilection, LCA usually occurs in adults and appears to be extremely rare in children.

Only a handful of pediatric cases have been reported thus far.[1,3-5] LCA is usually asymptomatic and is only discovered incidentally. Clinically, LCA may be characterized as a solid mass occurring most often along

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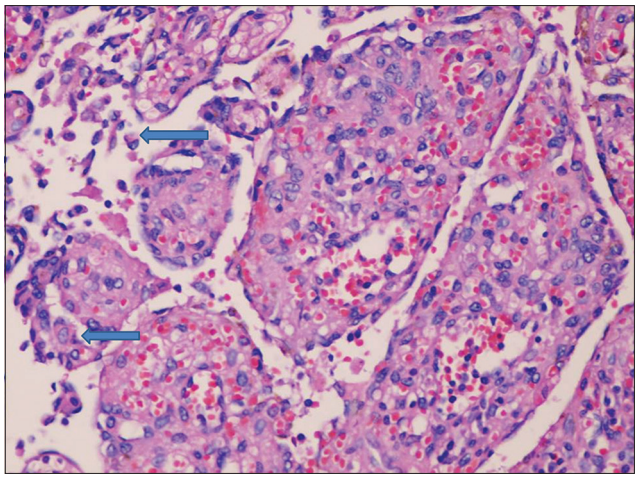

Figure 1: Spleen showing hypertrophied, dilated and tortuous vascular spaces are lined by plump cells (marked with arrow) with oval to indented nuclei with the appearances of sinus lining ("littoral") cells (H and E, ×400)

with splenomegaly, thrombocytopenia, or anemia, as well as a fever of unknown origin. The symptoms of anemia, thrombocytopenia, and abdominal pain may well be associated with hypersplenism. Grossly, there are two forms of LCA; diffuse multinodular and solitary (rare). The lesion can then be visualized on computed tomography (CT) scan, but the CT features of LCA are most often nonspecific. Like most vascular tumors of the spleen, the LCA shows changes in density during delayed image testing. Sonographic characteristics of LCA are not specific and include lobular splenomegaly with a diffuse, coarse, heterogeneous echotexture of the spleen, or a single echogenic mass that may contain cystic areas.[3-6] Little is known about the potential for malignant transformation in LCA.

To date, two subtypes of LCA with malignant potential have been described: Littoral cell angiosarcoma and littoral cell hemangioendothelioma. Patients affected by either may present with metastatic disease, even after undergoing a splenectomy. Peckova *et al*. have noted LCA to be frequently associated with internal malignancies in adults.[7]

Histological evaluation is likely to further the diagnosis by revealing a narrowed channel interspersed with large spaces that are lined by cuboidal cells, which contain hemosiderin deposits and/or hyaline globules. Atypical cells are absent, and mitotic activity is usually very low. Immunohistochemical stains reveal positive immunoreactivity to both the endothelial marker factor CD31 and the histiocytic marker CD68.[5,8]

In our case, histopathology showed a similar picture and is depicted in Figure 1.

Conclusion

LCA is rare in pediatric population and should be kept in mind when a child with thrombocytopenia develops splenic hematoma. Histopathology of the expunged tissue is the only diagnostic test in today's era of advanced radiology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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