Splenic Hamartoma: Immunohistochemical Profile

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Received: 03 Nov. 2015; Accepted: 06 Mar. 2016

Abstract- Splenic hamartoma is an unusual accidental finding with non-neoplastic feature that may be incidentally diagnosed because of its compression effect on surrounding organs. The predominant pathological feature leading diagnosis is circumscribed, un-encapsulated bulging nodules with focal fibrosis or cyst. The histopathological feature of splenic hamartoma is the positivity for CD8, and occasionally for CD31, CD34, and CD68 biomarkers on vascular wall but the cells are frequently negative for CD21. Herein, we describe a case with initial diagnosis of nephrolithiasis due to abdominal pain that was finally confirmed as splenic hamartoma by histopathological assessments and detection of CD8, CD31, CD34, and CD68 positivity.

Keywords: Splenic hamartoma; Immunohistochemistry

Introduction

The most frequent benign mass lesions of the spleen include hemangioma, cysts, and hamartoma that the latter has been rarely reported in the literature (1). Since 1961 that hamartoma was firstly described by Rokitansky, some researchers attempted to find out the pathological and imaging details of this tumor (2). Epidemiologically, splenic hamartoma can affect both genders similarly and may be appeared in all age groups predominantly in older adults (3). Because of asymptomatic feature in initial stages, this tumor may be detected incidentally. Pathologically, splenic hamartoma is characterized by a tumor-like proliferation of the tissues of normal spleen (4). The main pathological characteristics of splenic hamartoma include well-circumscribed, unencapsulated bulging nodular lesions adjacent to normal parenchyma of the spleen that may be also surrounded by fibrosis (5). Because this feature may be discovered in other common lesions such as hemangioma, lymphangioma or other vascular lesions, relying on specific parameters may facilitate differentiating splenic hamartoma as a very rare phenomenon of other more common masses. In this regard, positivity of CD8 marker has been shown to be a specific marker for detection of hamartoma (6). Moreover, applying imaging tools such as sonography, computed tomography (CT), or magnetic resonance imaging (MRI) can help the clinicians to accurately diagnose of the tumor (7). Herein, we describe a case with initial diagnosis of nephrolithiasis due to abdominal pain that was finally confirmed as splenic hamartoma by histopathological assessments.

Case Report

The case presented was a 47-year-old with the previous history of flank pain that diagnosed as nephrolithiasis prior to referring our hospital. He referred to our center for further evaluation of a splenic mass found incidentally. Abdominal ultrasound found a solid, slightly hyperechoic mass in the spleen with 16×12×5 cm³ in size. The patient was candidate for splenectomy. After the operation, the specimen was sent to our pathology laboratory for histopathological examination. The gross examination showed a mass with the dimension of 16×12×5 cm³, weighing 300 g and containing a 6 cm well-defined cream-brown mass in the subcapsular area. Using routine hematoxylin and eosin (H and E) staining, it was revealed a neoplasm exclusively composed of red pulps elements without intervening well-formed white pulps. No necrosis or mitosis is identified. A mild level of hemorrhage and degenerated changes were also found. Immunohistochemistry study was scheduled to detect the biomarkers of CD31, CD8, CD68, CD34 and CD21...
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on infiltrative cells and vessels wall revealing positivity for CD31, CD8, CD68, and CD34 biomarkers (Figure 1). In final, the diagnosis of splenic hamartoma was confirmed by light microscopic assessment.

Discussion

Splenic hamartoma is an unusual accidental finding with non-neoplastic feature. This mass is commonly observed in adults but about 20% of lesions can be detectible within childhood. A wide incidence range of splenic hamartoma has been reported in the literature from 0.015% to 2.7% (8). The presenting symptoms of the tumor are directly related to the size and in fact to its compression effect on surrounding organs. The most common focal symptoms of the tumor include nonspecific abdominal and flank pain with a feeling of a bulging on the left side. Hematological findings are not common however a degree of anemia and thrombocytopenia may be detected (5). As previously pointed, the predominant pathological feature leading final diagnosis is circumscribed, un-encapsulated bulging nodules with focal fibrosis and cyst. The size of the lesion widely varies between a few millimeters and as large as 20 cm (9). The prevailing histological finding includes vascular channels lined by slightly plump endothelial cells without atypia, mixed with intervening splenic red pulp-like stroma with or without white pulp (10). Other less common pathological findings include extramedullary hematopoiesis with elevated numbers of eosinophils, macrophages, and mast cells. The prominent histopathological feature of splenic hamartoma is the positivity for CD8 biomarkers on vascular wall. Along with this marker, other CD markers including CD31, CD34, and CD68 may be also positive, but the cells are frequently negative for CD21 (11,12). These biomarkers have a key role in differentiation of splenic hamartoma from other differential diagnoses such as hemangiomas (that is commonly negative for CD8) (10), Littoral cell angioma (that is highly positive for CD21) (11), and Lymphangioma (with CD8 negativity) (13).

As shown in our report, the case was incidentally diagnosed due to initial ultrasonography because of flank pain due to nephrolithiasis. Further histopathological assessment followed by immunohistochemistry findings reached to the diagnosis of splenic hamartoma. The detection of CD8, CD31, CD8, CD68, and CD34 positivity and CD21 negativity could confirm the final diagnosis.

Figure 1. IHC study revealed diffuse positive in infiltrative cells and vessels wall for IHC study with CD 34, CD31 markers and negative for CD21(×200)

References