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Case Report

Myoid Angioendothelioma of the Spleen: A Rare Case Report and Literature Review

Bita Geramizadeh[1,2], MD;
Mohammadhossein Anbardar[1], MD;
Seyed-Ali Malekhosseini[3], MD

[1]Department of Pathology, School of Medicine, Shiraz University of Medical Sciences, Shiraz, Iran;
[2]Transplant Research Center, Shiraz University of Medical Sciences, Shiraz, Iran;
[3]Department of Surgery and Transplantation, Shiraz University of Medical Sciences, Shiraz, Iran

Correspondence:
Bita Geramizadeh, MD;
Transplant Research Center,
Department of Pathology,
Nemazee Hospital,
P.O. BOX: 71345-1864
Shiraz, Iran
Tel\Fax: +98 71 36473954
Email: geramib@gmail.com
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Abstract

Most common tumors of the spleen are hematologic and lymphoid malignancies. Non-lymphoid and non-hematologic tumors of the spleen are very rare, the most common of which are vascular tumors. This group of tumors in the spleen is composed of heterogeneous tumors such as hemangioma, angioendothelioma (AE), littoral cell angioma, and angiosarcoma. There are several histologic forms of AE such as epithelioid AE, Kaposiform AE, and myoid AE. Among these splenic vascular tumors, myoid angioendothelioma (MAE) seems to be the least common type. It is a distinct tumor; composed of endothelium-derived tumor cells and a special type of stromal tumor cells that seems to have borderline low-grade malignant potential. Herein, we report our experience with a young woman presented with chronic abdominal pain and splenic mass that turned out to be myoid AE of spleen with an uneventful surgery and excellent recovery period. To the best of our knowledge, such an occurrence is very rare in the spleen.

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Keywords ● Spleen ● Malignancy ● Myoid angioendothelioma

What's Known

- Angioendothelioma of the spleen is a rare tumor with several subtypes. The least common type is myoid angioendothelioma.

What's New

- Myoid angioendothelioma is a rare disease that was seen in a young woman with vague symptoms. Complete excision can be curative with no recurrence and complication.

Introduction

Vascular neoplasms are the most common primary nonlymphoid, nonhematopoietic neoplasms of the spleen, including hemangioma, lymphangioma, hamartoma, littoral cell angioma, angioendothelioma, and angiosarcoma.[1,2] There are different types of splenic angioendothelioma such as kaposiform angioendothelioma, myoid angioendothelioma (MAE) and epithelioid angioendothelioma.[2-4] The age of presentation, pathologic findings, and clinical behavior are different in these types of AE.[1]

In this report, we will describe our experience with a rare case of splenic MAE (with the patient's permission and consent).

Case Report

A 38-year-old woman presented with epigastric abdominal pain and fullness since several months. Her past medical and family histories were unremarkable. On physical examination, no organomegaly was detected. Complete blood count values were all normal. Biochemical laboratory findings were also unremarkable. Abdominal sonography showed a hypoechoic mass in the spleen. A computed tomography (CT) scan revealed a mass in the splenic parenchyma measuring 3×3 cm

(figure 1). Fine-needle aspiration cytology of the mass was inconclusive and showed very few small clusters of spindle cells, reported as unsatisfactory. Tru-cut biopsy showed a hypervascular tumor with tiny and small vascular channels with no significant atypia or mitosis. No interconnecting or anastomosing blood vessel was seen. Immunohistochemistry (IHC) was performed which showed reactive tumors cells with endothelial markers such as CD31 and CD34, nonreactive for CD45, and cytokeratin. Proliferative index by Ki67 was very low (less than 1 per 10 HPF). Therefore, with the diagnosis of splenic vascular tumor, most probably AE, the patient underwent splenectomy.

The resected spleen was 18×10×5 cm with smooth external surface and intact capsule. Moreover, serial cut sections showed a well-defined round, pinkish and spongy mass measuring 3×3×3 cm (figure 2). Histologically, the same as tru-cut biopsy, the lesions showed a well-defined mesenchymal tumor mainly composed of spindle to histiocytic tumoral cells, which formed small capillary-like vascular channels. These tumor cells were embedded in eosinophilic stroma rich in spindle to plump stroma cells. Tumor cells were reactive for CD31 (figure 3a), with low MIB-1 index and negative leukocyte common antigen (LCA). The stromal cells were reactive with smooth muscle actin (SMA) (figure 3b). There was little nuclear atypia and the mitotic rate was low (about 1/10 high-power field). No anastomosing or cavernous channels were present. No necrosis was identified.

According to the histological and immunohistochemical findings, diagnosis of MAE was confirmed in the splenectomy specimen. Because of the small size of the tumor, complete excision was performed without any further medical treatment. However, she is currently under regular follow-up.

The patient had an uneventful postoperative period and she was discharged in a good general condition. Now, after about 3 months, she is completely well.

Discussion

The spleen is composed of two compartments, namely red pulp and white pulp. Most of the splenic tumors originate from white pulp (i.e. lymphohematopietic tumors). The second most common tumors of the spleen are vascular tumors originated from red pulp.[1]

Angioendothelioma is a vascular tumor with morphologic appearance and a clinical course intermediate between hemangioma and angiosarcoma. This tumor is most commonly seen in the soft tissue and subcutaneous tissue.[2] However, this tumor can also be seen in the liver, lung and other visceral organs, but the behavior and clinical characteristics of the tumor in these two locations are completely different.[2] This tumor is very rare in the spleen.[3] Splenic angioendothelioma has a wide range of morphologic and biologic behavior, composed of kaposiform, myoid and epithelioid angioendothelioma.[3,4] Among these types, MAE is the least common and 6 cases have been reported in the English literature so far.[5-7]

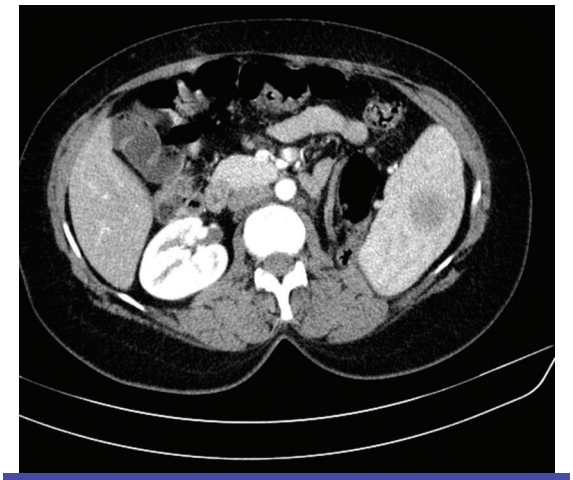

Figure 1: CT scan of the abdomen shows a 3 cm splenic mass (note congenital absence of the left kidney).

Figure 2: Gross of the spleen shows a well-defined 3 cm mass.

Table 1 shows the clinicopathologic and radiologic findings in these three cases.

The age of presentation was from 3 to 46 years with female to male ratio of 4/3, including our case.[4,5] Most of the previous cases were detected incidentally during investigation for other underlying diseases and only one case was found secondary to patient's evaluation for the cause of abdominal pain and discomfort.[5]

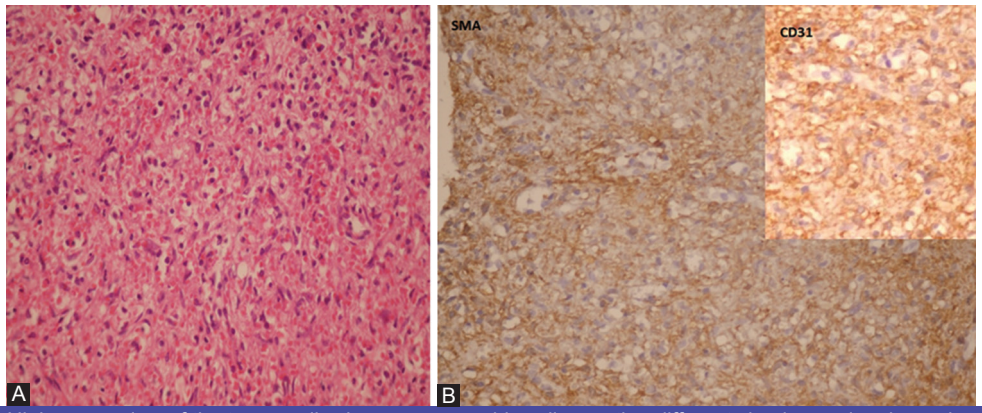

Figure 3: A) High power view of the tumor cells show monomorphic cells creating different sized vascular channels accompanied with stromal cells. (H&E, ×400). B) Sections from immunohistochemistry of the tumor show reactivity of the stromal cells with SMA. The inset shows positive CD31.

Table 1: A comparison of the clinical characteristics of the reported 6 cases and the current case of myoid angioendothelioma of the spleen

Author	Year	Age (year)	Sex	Accompanying disease	Presenting symptoms	Size of (cm)	Prognosis
Jang et al.[1]	2013	41	F	Rectal cancer	Incidental	3	Alive
Chan et al.[5]	2005	4	M	Wilms' tumor	Incidental	2.2	Alive
Karim et al.[6]	2004	51	M	Abnormal liver function tests	Incidental	3.5	Alive
Kraus et al.[7]	1999	3	F	Beckwith- Wiedemann syndrome	Incidental	2.5	Alive
		7	M	None	Abdominal discomfort	4	Alive
		43	F	Pancreatic mucinous neoplasm	Incidental	1.9	Alive
Current case	2015	36	F	None	Abdominal pain	3	Alive

Imaging in MAE of the spleen can be very helpful, but in contrast to hemangioma, it is hypoechoic in ultrasonography (US) with clear demarcation. CT scan shows low vascularity, which is completely different from hemangioma.[6] The gold standard for the diagnosis of splenic MAE is pathology. The lesions classically show a range of morphologic features with epithelioid to spindle-shaped tumoral cells forming small vascular channels, mild atypia, low mitotic figures, and lack of necrosis.[2,3] This tumor displays intermediate biologic behavior, although until now the number of cases has been too low to make a definite evaluation of the prognosis and survival of patients with MAE. However, metastasis or aggressive behavior has not been reported among the previous cases.[5,6]

Splenic MAE is such a rare tumor that, to the best of our knowledge, only 6 cases are reported in the English literature. These tumors, as in our case, are mainly composed of spindle and epithelioid tumoral cells and the endothelial nature of cells has been confirmed by characteristic immunoreactivity for CD31, CD34, and factor VIII-related antigen.[4] Intervascular stroma contains bland intermediate sized polygonal to spindle cells with little atypia and low mitosis, and these cells show strong reactivity for SMA and actin, but negative for endothelial markers.[8-10] Our case showed no reactivity for SMA in tumoral cells and most of the cells were reactive for endothelial markers, and stromal cells were strongly reactive with SMA.

Definitive diagnosis of MAE is based on morphological and immunohistochemical findings that differentiate it from other vascular lesions of the spleen, including splenic hemangioma, lymphangioma, angiosarcoma, littoral cell angioma, hamartoma, and sclerosing angiomatoid nodular transformation (SANT) of the spleen.

Splenic hemangioma is the most common primary tumor of the spleen, often of cavernous variety. Grossly appear as circumscribed, nonencapsulated, honeycomb-like mass and histologically consisting of large interconnected blood-filled spaces lined by a monolayer of bland looking endothelial cells that differentiate it from MAE.[1,2,7]

Splenic lymphangioma is composed of thin-walled cystic structures of varying size lined by flat, bland-looking endothelium and filled with pink proteinaceous material devoid of red blood cells.[1,2,7] Littoral cell angioma is

a unique lesion to the spleen. Grossly, the tumor is multiple, nodular, and dark-red with spongy cystic appearance. Microscopically, it is characterized by anastomosing and cyst-like vascular channels with papillary projections lined by sloughed plump endothelial cells often showing hemophagocytosis. The tumor cells demonstrate dual expression of endothelial and histiocytic markers that differentiate it from other vascular tumors. The immune phenotype has been established to be $CD68^+/CD31^+/Lysosyme^+/vWF^+/CD21^+/CD34^-/CD8^-$, which is indicative of dual derivation of the neoplasm.[1,2,7] The characteristic morphology includes anastomosing and cyst-like vascular channels with papillary projections lined by sloughed plump endothelial cells and immunoreactivity for CD68.

Primary splenic angiosarcoma is rare and clinically aggressive tumors with very poor prognosis. Grossly, the lesions are variegated with multiple foci of hemorrhage and necrosis. Histologically, there are anastomosing networks of blood vessels with highly atypical spindled cells that show enlarged, irregular hyperchromatic nuclei. Mitotic figures are prominent and necrosis is present. The cells are immunoreactive for endothelial markers with high proliferative index.[1,2,7] Splenic hamartoma is the term used for a nodular lesion of the spleen, composed of unorganized vascular channels of varying width, with intervening red pulp-like disorganized stroma with or without lymphoid follicles. The endothelial cells are similar to those of normal splenic sinuses. Although rendering a diagnosis can be difficult, endothelial cells that are positive for CD8 are a key feature that differentiates hamartoma from other vascular lesions of the spleen.[1,7]

SANT is a recently identified nonneoplastic vascular lesion of the spleen. Microscopically, it consists of multiple well-circumscribed vascular nodules showing plump endothelial cell and extravasated erythrocytes. The nodules are encircled by a variable lymphoplasmacytic infiltrate, spindle cells, and collagenous stroma. The vascular nodules display a complex mixture of endothelial phenotypes resembling splenic sinusoids ($CD34^-/CD31^+/CD8^+$), capillaries ($CD34^+/CD31^+/CD8^-$), and small veins ($CD34^-/CD31^+/CD8^-$). It has been supposed that SANT represents an especial hamartomatous transformation of splenic red pulp in response to an exaggerated nonneoplastic stromal proliferation.[11,12] The nodular pattern of growth, presence of collagenous stroma between the nodules, and presence of three distinct types of vessels according to immunostaining can differentiate it from other splenic vascular tumors.

Another important task of the pathologist is subtyping of the different types of angioendothelioma. Epithelioid hemangioendothelioma of the spleen is very similar to its liver counterpart (i.e. composed of epithelioid cells creating small vascular channels and with no myoid SMA positive stromal cells).[8,9,13]

Another angioendothelioma in the spleen is kaposiform angioendothelioma; consist of irregular and infiltrating nodules of densely packed spindle-shaped tumor cells with small slit-like blood vessels, which were separated with hyalinized hypocellular fibrous stroma. They are almost exclusively seen in pediatric age group and are extremely rare in adults. It means that vascular channels are very similar to Kaposi sarcoma (i.e. slit-like).[10,14,15]

The therapeutic strategies for splenic myoid angioendothelioma have been limited and the only effective therapy is complete splenectomy.[5]

Conclusion

MAE is a rare splenic vascular tumor that seems to have an indolent course. Although the amount of literature regarding this tumor is very low, however, it seems that complete excision is a complete cure for this type of angioendothelioma.

Conflict of Interest: None declared.

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