Heterotopic Ovarian Splenoma

Report of a First Case

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We present a case of ovarian splenoma, a form of heterotopic splenic hamartoma consisting of red pulp tissue. The hamartoma was located in ovarian stroma in an otherwise normal ovary. The histology showed interanastomosing vascular channels of splenic sinusoidal red pulp lined by cells that were immunoreactive for antibodies to von Willebrand antigen and CD8, findings consistent with splenic lining cells. The sinuses were lined by cuboidal to flattened cells with ovoid and grooved bland-looking nuclei. Ultrastructurally, the tumor cells showed Weibel-Palade bodies and lysosomes. To our knowledge, this is the first case of splenic tissue arising in an ovary, and it underlines the trend noted in the literature that splenic hamartoma, although a rare entity, can arise in many retroperitoneal organs, including the ovary.

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Splenoma (splenic hamartoma) is a rare, benign splenic vascular lesion derived from splenic sinus-lining cells. It is characterized by the exclusive presence of red pulp tissue without associated white pulp, although there may be scattered lymphoid cells. Heterotopic splenic tissue is well known to occur in retroperitoneal organs such as the retroperitoneal soft tissue, pelvis, testis, and pancreas.1–3 These lesions are usually small incidental findings. When they are sizeable and located in the spleen, they may occasionally cause thrombocytopenia secondary to platelet sequestration.4 Although more than 140 splenic hamartomas have been described in the spleen5 since their initial description by Rokitansky in 1861, this lesion has never been reported in an ovary. We report what we believe to be the first such case.

REPORT OF A CASE

This patient was a 45-year-old white woman who had a long history of menometrorrhagia and known uterine fibroids. In the recent past, she had experienced heavy menses with passage of clots, and she had occasionally required transfusion. More recently, she had experienced severe daily vaginal bleeding. Her endometrial biopsy was consistent with the presence of anovulatory cycles. Despite multiple regimens of hormones and medical therapy, her symptoms did not improve. The patient also had a past history of multiple medical problems that included chronic renal failure (for which she was on hemodialysis), chronic hypertension, type 2 diabetes mellitus, and a history of pulmonary embolism. There was no history of abdominal trauma. She underwent a total vaginal hysterectomy and right salpingo-oophorectomy for definitive therapy of her menometrorrhagia. The surgeon did not encounter any fibrous connecting band between the ovary and the spleen.

PATHOLOGIC FINDINGS

Grossly, the uterus was enlarged, weighing 274 g, and it contained multiple intramural leiomyomas. The ovary was slightly enlarged, measuring $4 \times 2.5 \times 3$ cm. The external surface of the ovary was unremarkable. The cut surface revealed a soft, dark red, well-circumscribed nodule measuring 1.5 cm in the stroma of the ovarian cortex. There were also a few cortical cysts lined by smooth glistering walls. The remaining ovarian parenchyma appeared unremarkable.

Microscopically, the reddish nodule consisted of a small, nonencapsulated, well-defined sinusoidal and vascular lesion within the ovarian cortex that did not compress the adjacent ovarian tissue (Figure 1). The anastomosing and disorganized vascular sinusoidal channels produced irregular blood-filled lumina lined by tall and plump endothelial-like cells that contained vesicular nuclei and small nucleoli. These lining cells occasionally demonstrated central grooves. No cytological atypia or mitotic figures were identified. Other lining cells contained red cell fragments and evidence of erythropagocytosis (Figure 2), and scattered erythroblasts were found in some of the sinusoidal lumens, a finding in support of extramedullary erythropoiesis. Reticulin and periodic acid–Schiff stains highlighted the sinusoidal nature of the lesion. These stains also demonstrated rare ring fibers, delineating a characteristic of splenic sinuses (not shown). Immunohistochemical stains were performed to evaluate the nature of the lining cells. They showed diffuse and strong staining for von Willebrand antigen (factor VIII:Ag; not shown) and also expressed immunoreactivity for CD8 (T cells, splenic lining cells; Figure 3, A). Scattered CD68 (KP-1, macrophages)–positive cells are also seen (Figure 3, B), especially in the histiocyte-rich areas analogous to the cords of Bilroth. A CD21 immunostaining was performed for the presence of CD21-positive dendritic cells in aggregate (seen in accessory spleen) with negative results. Ultrastructurally, the cytoplasm of the lining cells clearly demonstrated the presence of Weibel-Palade bodies, lysosomes, and ingested debris (Figure 4).

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The hamartoma in this case had the same histopathologic and immunohistochemical features as described when hamartomas arise in the spleen. A splenic hamartoma or splenoma is a well-circumscribed red pulp-only lesion that consists of anastomosing sinusoidal channels containing irregular lumina and cystlike spaces. The lining endothelial-like cells appear tall and plump, with large vesicular nuclei and small nucleoli. We believe that the classic histologic features, along with the typical immunoreactivity for factor VIII–related antigen and CD8 of splenic lining cells, are in support of a splenic origin. Ultrastructural characteristics of splenic lining cells, including the presence of Weibel-Palade bodies, lysosomes, dense bodies, and phagocytosed material in the cytoplasm, also support the diagnosis.

Because of its histologic similarity to other vascular lesions, the differential diagnosis may include cavernous and capillary hemangioma, bacillary angiomatosis, and other rare vascular lesions. The spongelike anastomosing sinusoidal architecture separated by scant connective tissue stroma can easily be dismissed as a hemangioma. However, the findings of slitlike spaces and absent cavernous vascular spaces filled with red blood cells and lined by thick fibrous bands are not features of cavernous hemangioma. Pyogenic granuloma or capillary hemangioma, which pyogenic granuloma resembles, shows a lobular configuration of granulation tissue sprinkled with inflammatory cells and sprouting capillaries grouped around a parent vessel; these features were not seen in this case. Masson’s hemangioma, or intravascular papillary endothelial hyperplasia, is an exuberant intravascular reactive endothelial cell proliferation secondary to an organizing thrombus. The lesion in this case was not intravascular in location, and it lacked the papillary endothelial cells. A careful search and a periodic acid–Schiff stain for thrombi failed to reveal their presence. Angiosarcomas, unlike this present case, usually demonstrate a more haphazard growth pattern, solid tumor nests, cytologic atypia, and mitotic activity. All of these were lacking in this case.

The question of whether this lesion was congenitally present in the ovary or whether it was an acquired lesion is unresolved. A rare congenital splenic-gonadal fusion has also been described in an ovary. Cases described by Putschar and Manion were not limited to nondescended gonads and may or may not have had a fibrous band connecting the spleen and the gonadal-mesonephric remnant. Points arguing against this anomaly include the findings that the vast majority of cases of this type of fusion involve the left gonad (this case involved the right gonad) and that most cases reported are in males. We could not, however, rule out the possibility that this may be the discontinuous form described by Putschar and Manion. Because the surgeon did not encounter such a splenic-gonadal band in this case, this could also be regarded as an accessory spleen variant. However, the absence of CD21-positive dendritic cells in aggregates was not supportive of an accessory spleen.

We believe that this case represents a splenic hamartoma (splenoma), with its characteristic findings, albeit in an unusual, ectopic location in the ovary. It underscores the consideration that heterotopic splenoma must be included in a differential diagnosis of vascular lesions arising in the ovary.

Figure 1. Medium-power view of the splenic hamartoma showing a nonencapsulated, well-defined nodule composed of anastomosing, blood-filled, sinusoidal channels in haphazard arrangement and larger vessels with thicker walls (hematoxylin-eosin, original magnification ×250).

Figure 2. Disorganized and irregular sinuses mixed with cordlike elements. The vascular lumen is lined by tall and plump endothelial-like cells with vesicular nuclei and small nucleoli, some of which contain red cell fragments, an evidence of cytophagic activity (hematoxylin-eosin, original magnification ×500).

Figure 3. CD8 (A) and CD68 (B) immunostaining indicating typical immunoreactivity of splenic red pulp tissue. Note the sievelike arrangement typically seen in splenic red pulp (immunoperoxidase, original magnifications ×250 [A] and ×600 [B]).
Figure 4. Electron micrograph of the tall lining cells with discontinuous basement membrane and apical buds of cytoplasm showing the presence of Weibel-Palade body (inset), lysosomes, and ingested debris (original magnification ×7500).

References