Case report

Incidental splenic littoral cell angioma complicating a case of colon cancer: A case Report

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Abstract

Littoral cell angioma (LCA) is a recently described rare generally benign primary vascular neoplasm of the spleen originating from the lining cells of the splenic red pulp sinuses that is usually discovered incidentally. LCA may be associated with epithelial malignancies and may itself also have malignant potential. We report the case of a 71–year–old woman who presented with intraoperative bleeding from the spleen during sigmoidectomy for colonic adenocarcinoma. Histopathological examination of the removed spleen revealed multiple haemorrhagic lesions diagnosed as littoral cell angioma. This case has been reported due to its rarity and to highlight how its accidental detection, unique and unexpected presentation complicated a case of colonic carcinoma. Individuals diagnosed with this tumour must be carefully evaluated to exclude primary, secondary and synchronous malignancies.

Keywords

Incidental, splenic, littoral cell angioma, colon, cancer.

Introduction

The most common primary tumours of the spleen are vascular in origin and include hemangiomas and hamartomas. Littoral cell angioma (LCA) is a rare neoplasm that arises from the cells lining the splenic red pulp sinuses, first described by Falk et al in 1991[(1)]. The exact incidence of littoral cell angioma is unknown with approximately 80 cases being reported in the English literature since its original description. [(2,3)] LCA has been shown to exhibit malignant potential and may be associated with visceral malignancies. [(4)] These tumours are often underdiagnosed because of the limited number of cases, atypical imaging modalities and unclear etiopathogenesis. Here, we report a case of this rare vascular neoplasm with an unexpected presentation complicating a case of colon cancer.

Case description

A 71– year– old female, Iranian by origin, known to have dyslipidemia and a positive family history of breast cancer, leukemia and gastric cancer presented to the Surgical Outpatient Department with a month history of bloody diarrhea. Physical examination, routine hematological investigations and tumor markers were within normal limits. A computerized tomography scan of the abdomen showed a 6.5cm mass with irregular mural thickening and apple core appearance in the distal sigmoid colon. A PET–CT scan demonstrated a hypermetabolic lesion in the sigmoid colon and heterogeneous FDG activity in the whole skeleton. Colonoscopy revealed a 1.2 cm semipedunculated polyp at 50 cm and another large lobulated circumferential mass at 20 cm from the anal verge. An anterior resection of the sigmoid colon was performed. During surgery, the patient developed bleeding from the spleen that necessitated splenectomy. The histopathological examination of the sigmoid colon showed moderately differentiated

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Figure 1. Spleen with multiple haemorrhagic lesions

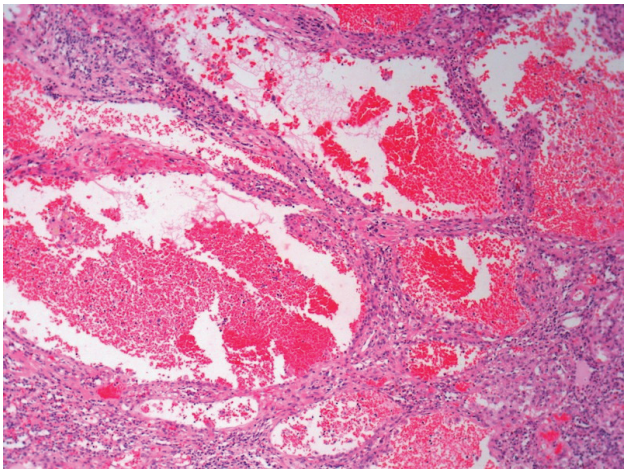

Figure 2. Splenic haemorrhagic lesions showing multiple dilated vascular spaces (H &E, x 200)

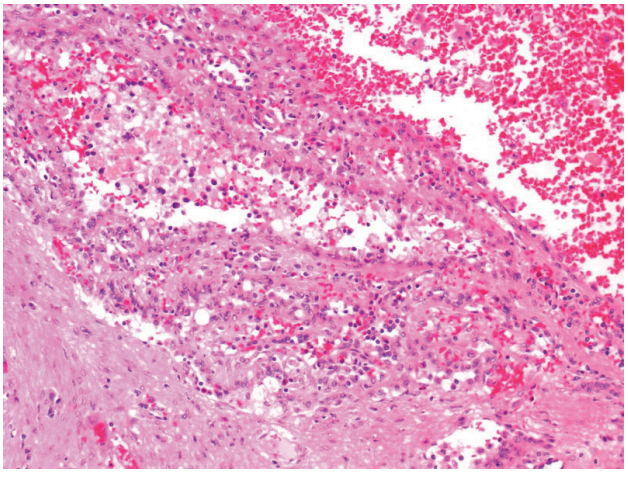

Figure 3. Vascular spaces with dissociated lining cells in the lumen (H &E, x 400)

adenocarcinoma invading the muscularis propria. The pericolic lymph nodes did not show evidence of metastatic adenocarcinoma. The splenectomy specimen was received in multiple fragments altogether weighing 186.5gm and it revealed multiple circumscribed hemorrhagic lesions measuring up to 1 cm in diameter (Figure 1). Microscopy of the splenic hemorrhagic areas revealed unencapsulated

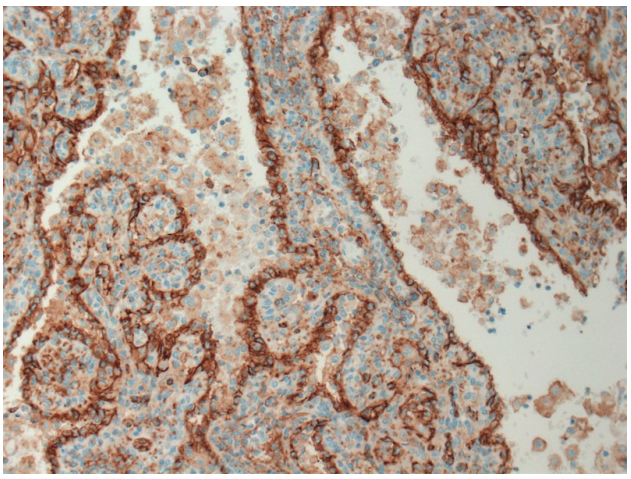

Figure 4. CD 31 Immunostain positivity in the lining cells of the vascular spaces (x 400)

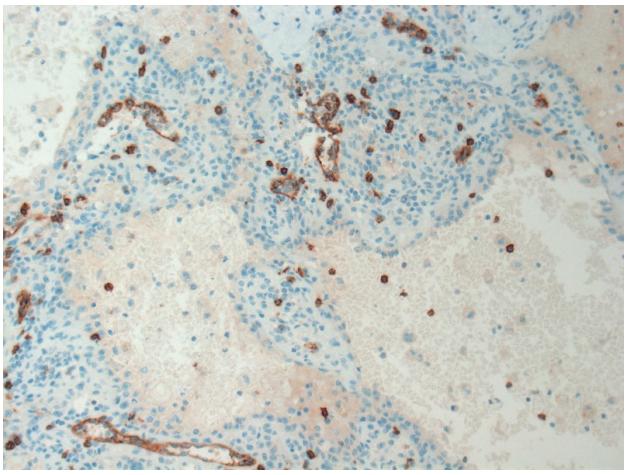

Figure 5. CD8 Immunostain negativity in the lining cells of the vascular spaces (x 400)

vascular spaces with focal pseudopapillary configuration (Figure 2). Dissociated cells were seen in the lumen (Figure 3). Immunohistochemistry showed that the lining cells are positive for CD31, FVIIIAg, CD68, S100, CD21 but negative for CD8 and CD34 (Figures 4 & 5). A final diagnosis of moderately differentiated adenocarcinoma, sigmoid colon (pT2N0Mx, MAC stage B1) and Littoral cell angioma of spleen was rendered. Postoperatively she had an episode of deep vein thrombosis in the right limb which was successfully treated. The patient is currently asymptomatic upon follow–up at 19 months postoperatively.

Discussion

LCA is a recently described primary vascular neoplasm of the spleen with no soft tissue counterpart. LCA has no age or sex predilection. This

lesion can be completely asymptomatic and may represent an incidental finding by imaging or present with relatively benign findings of hypersplenism such as splenomegaly, thrombocytopenia or anemia and constitutional symptoms such as fever, abdominal pain etc. [5] Our patient presented for the first time as intra–operative bleeding from the spleen necessitating splenectomy during the surgical resection of colon cancer. A similar presentation of LCA presenting as splenic rupture and hemoperitoneum was reported by Pilz et al. [6]

Our patient was undetected by the preoperative scans which is explained by the nonspecific and non–diagnostic radiological features of LCA. They are reported to possess variable features on grayscale sonography, color Doppler imaging as well as contrast enhanced sonography. The sonographic appearance of littoral cell angioma mainly depends on the type and number of tumor vessels. [7]

Grossly, these lesions usually appear as multiple nodular spongy blood filled lesions in 84% of cases and their size may range from 0.1cm to 11cm in diameter, similar to ours demonstrating multiple dark brown splenic hemorrhagic lesions, the largest measuring 1 cm in diameter. The solitary presentation of splenic littoral cell angioma is very rare and occasionally these lesions can appear white on gross pathology. The spleen itself may appear grossly enlarged or, as was true for our patient look otherwise unremarkable. The diagnosis is usually confirmed by histopathological examination with its unique histological and immunohistochemical features which differentiates it from traditional cavernous and capillary hemangioma.

In general, the course of LCA is benign as supported here, however two subtypes of Littoral cell angioma with malignant potential have been described as "littoral cell angiosarcoma" and "littoral cell hemangioendothelioma". These littoral cell angioma variants show features consistent with littoral cell angioma histopathology as well as abnormal architecture, nuclear atypia, and necrosis. These variants may present with distant metastasis several months after surgery.

The association of LCA with visceral epithelial malignancies such as colorectal, thyroid, renal, pancreatic, ovarian and testicular cancers is documented in the literature and the most common malignancy being colorectal adenocarcinoma, such as in our patient. [4,8] The occurrence of these lesions in autoimmune disorders like Crohn's disease and metabolic diseases such as Gaucher's disease have also been reported. [9]

The etiology and histopathogenesis of LCA remains unclear. In cases associated with autoimmune diseases and visceral malignancies, underlying chronic infection and immune dysregulation has been postulated as factors triggering the development of these tumors. Immunohistochemically, the lining cells of LCA showed pattern similar to the dual differentiation potential of the reticuloendothelial cells lining the splenic sinus. [1] However the surprising CD8 negativity of these cells, as is in our patient, has raised suspicion regarding its origin from littoral cells. [10]

Symptomatic LCA are often treated by splenectomy and given the association of littoral cell angioma with other malignancies and reported cases of metastasizing littoral cell angioma, splenectomy is both diagnostic and therapeutic. Long term surveillance for the development of synchronous and metachronous lesions is required.

Conclusion

This report highlighted the importance of a stringent search for any other synchronous silent lesions elsewhere in known cases of malignancy that can otherwise cause unexpected complications. And even though rare, LCA should be included in the differential diagnosis of patients with multiple splenic nodules and history of malignancy.

References

1. Falk S, Stutte HJ, Frizzera G. Littoral cell angioma–novel splenic vascular lesion demonstrating histocytic differentiation. Am J Surg Pathol 1991; 15(11):1023–33.
2. Hu ZQ, AYG, Sun QM, Li W, Li L. The splenic Littoral cell angioma in China: a case report and review. World J Surg Oncol. 2011; 9:168.
3. Tee M, Vos P, Zetler P, Wiseman SM. Incidental littoral cell angioma of spleen. World J Surg Oncol. 2008; 6: 87.
4. Bisceglia M, Sickel JZ, Giangaspero F, Gomes V, Amini M, Michal M. Littoral cell angioma of the spleen: an additional report of four cases with emphasis on association with visceral organ cancers. Tumori 1998; 84: 595–9.

5. Harmon RL, Cerruto CA, Scheckner A: Littoral cell angioma: a case report and review. Curr Surg 2006; 63(5):345–50.

6. Pilz JB, Sperschneider T, Lutz T, Loosli B, Maurer CA. Littoral cell angioma in main and accessory intrapancreatic spleen presenting as splenic rupture. Am J Surg 2011; 201(2): e15–7.

7. Zhang YH, Liu LM, Wang WP, Ding H, Wang XN, Xia HS .Littoral cell angioma of the spleen Sonographic–Pathologic Comparsion. J Ultrasound Med 2013; 32(4):691–7.

8. Hansen T, Habekost M, Flieger D, Kirkpatrick CJ. Littoral cell angioma of the spleen. Association with colon and hepatocellular carcinoma. Pathologe 2010; 31(4):290–2.

9. Gupta MK, Levin M, Aguilera NS, Pastores GM. Littoral cell angioma of the spleen in a patient with Gaucher disease. Am J Hematol .2001; 68(1): 61–2.

10. Arber DA, Strickler JG, Chen YY, Weiss LM. Splenic vascular tumors; A Histologic, Immunophenotypic, and Virologic Study. Am J Surg Pathol.1997; 21(7):827–35.