



Case Report

Case Report: Littoral Cell Angioma in a Patient with Rectal Carcinoma

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Abstract

We present the case of the incidental finding of multifocal splenic nodular masses in a computed tomography scan performed in the context of the tumor follow-up after resection of a rectal carcinoma. The radiologic appearance suggested at first a hamartoma of the spleen, which showed to be growing over time. An elective splenectomy was performed after 4 years of watchful-waiting with regular radiologic controls (standard follow-up for the rectal cancer) and the histopathologic analysis revealed a littoral cell angioma.

Introduction

Etiologies for multifocal splenic lesions include infectious (abscesses, fungal disease), inflammatory processes, primary vascular and lymphoid neoplasms, metastatic disease, vascular processes (splenic infarcts in patients with portal hypertension or pancreatitis, emboli), and systemic diseases (myelofibrosis, lymphoma, leukemia, infections such as HIV/AIDS, mononucleosis). They can be observed in a very large variety of clinical settings, from asymptomatic to critically ill patients [1]. The etiology of a splenic neoplasia can be primary or secondary. The spleen is in principle an infrequent site of tumor metastasis, but metastasis can arise from breast, lung, ovary, colon cancer and skin melanoma [1]. Splenic metastases are seen in 2%–9% of patients with metastatic end-stage cancer [2]. Primary neoplasms involving the spleen can be divided into lymphoid neoplasms, which primarily arise from the white pulp and vascular neoplasms, which primarily arise from the red pulp [3]. Vascular tumors are the most common primary neoplasm of the spleen. Most of these lesions are benign hemangiomas, which derive from vascular endothelium. Malignant vascular tumors such as hemangioendotheliomas and hemangio-sarcomas are much rarer. Lesions derived from lymph vessel endothelium (lymphangiomas and lymphangiosarcomas) may also occur, as well as vascular hamartomas of the spleen, which are tumors of the pulp cord [4]. The littoral cell angioma

has first been described by Falk and al. in 1991 as a neoplasia originating from splenic sinus lining cells (are therefore always situated in the red pulp). Therefore, they express a typical immunohistochemical panel that shows the distinct hybrid endothelial-histiocytic phenotype of littoral cells. This neoplasia appears to be unique to the spleen, without any soft tissue or lymph node counterpart [1]. Most described cases of littoral cell angiomas are multifocal [4,5], and can also involve accessory organs such as the pancreas [6] or the liver [4]. Disseminated disease is reported in rare cases and is a sign of atypical but not frankly malignant course of this entity [7]. According to a new study comprising the analysis of 435 cases of Littoral Cell Angioma, this entity should be considered as a benign vascular tumor, which is the intrasplenic manifestation of abnormal body function (due to the high rate of coexisting malignancy or immune dysfunction) [8].

Case Description

The 58y old male patient first presented in April 2018 at the emergency department of our hospital with a high-grade suspicion of rectal carcinoma after a colonoscopy. Except for irregularity in defecation habits since 9 months, the abdominal anamnesis was unremarkable. Relevant to mention are a daily nicotine (50 PY) and alcohol (2 Standard Units /d) consumption. The abdomen was clinically inconspicuous. The digital rectal examination

showed traces of blood as expected in a case of rectal cancer. The laboratory results identified a light inflammation with Lc 6.2 G/L and CRP 27.4 mg/L. The rest of the values were within the standard ranges and especially showed no signs of hypersplenism (no anemia and no thrombocytopenia). A computed tomography scan was performed, which confirmed the diagnosis of rectal cancer (pictures suggesting a Stade T4 N2), and detected a circular 2 cm lesion at the cranial pole of the spleen without contrast-uptake in the early phase. Therefore, the lesion was not suspect of a metastasis. The diagnostic of a hamartoma of the spleen was suggested. In the presence of a suspect liver lesion an additional MRI was performed, which excluded metastasis of the liver and showed the splenic lesion possibly corresponding to a hamartoma (Figures 1-3).

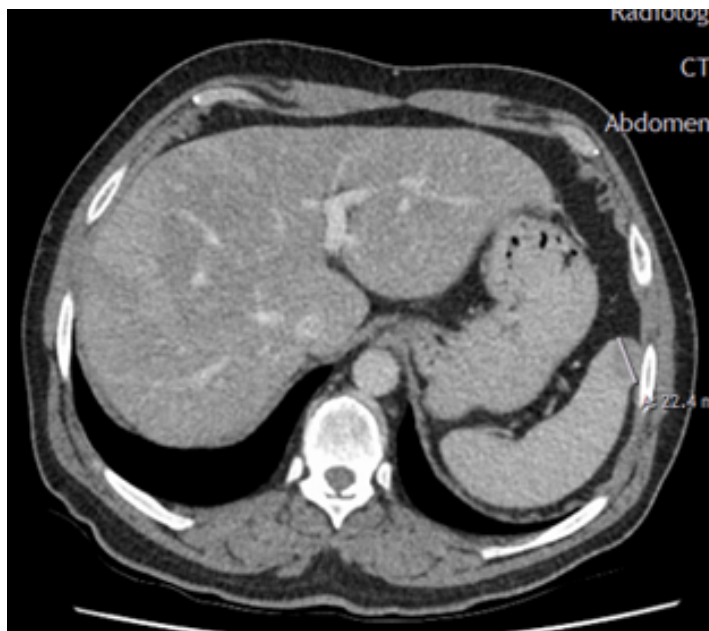


Figure 1: CT scan (April 2018) showing a 24 mm circular mass without increased contrast-uptake in the early phase.

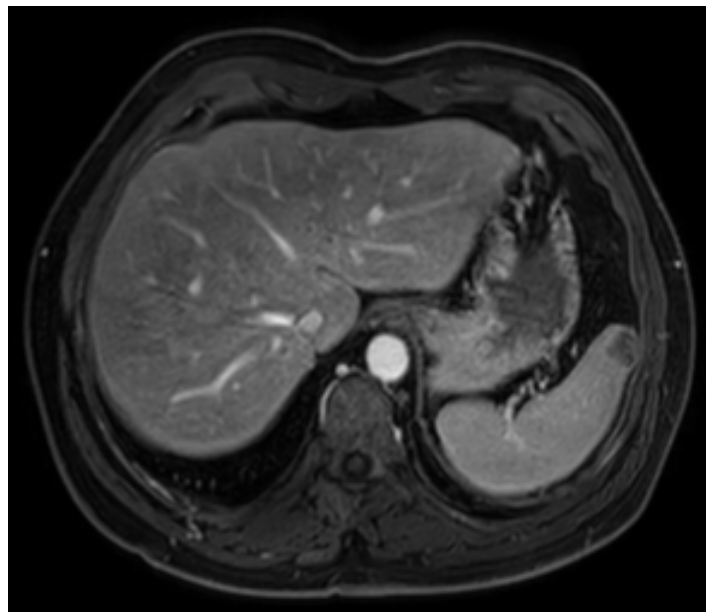


Figure 2: MRI (April 2018) showing the well-circumscribed lesion suspect of a hamartoma.

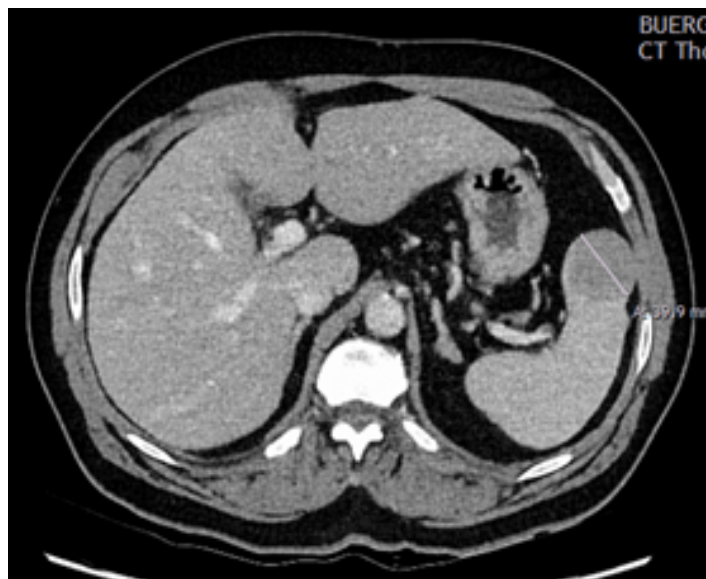
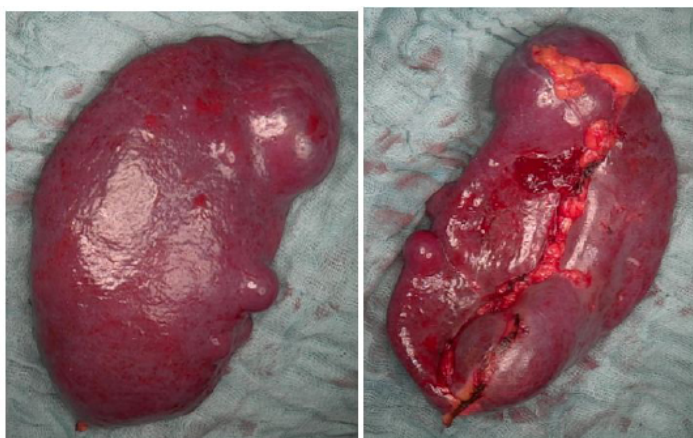


Figure 3: CT (December 2021) showing the enlargement of the lesion, measuring 36 mm.

The rectal cancer was treated in a curative-intention and in the course of the oncological follow-up, the spleen lesion appeared to grow over time. The last computed tomography scan (December 2021) showed an enlarged lesion, now 3.6 cm in diameter, as well as two new lesions of respectively 1.4 cm and 1.0 cm. The radiological examination also showed a broad-based abdominal incisional hernia without signs for incarceration. A hernia repair was recommended to avoid later complications and the patient decided to undergo the abdominal repair combined with a splenectomy as a one-time operation given the growing mass of unknown etiology. The operation was performed in April 2022 and showed a normal-sized spleen with three round lesions of various sizes and splenic aspect (Figures 4A,B).



Figures 4: Specimen after splenectomy: 13 x 10 x 6 cm, with the three round lesions.

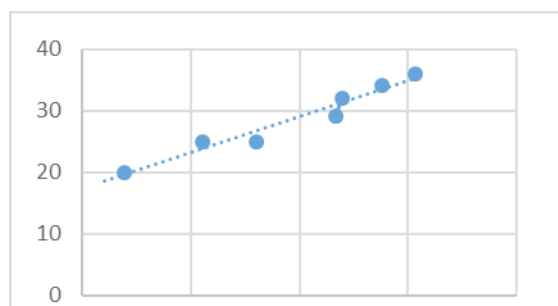
The spleen was sent for pathological analysis, which revealed the diagnosis of Littoral Cell Angioma. The case was discussed on our multidisciplinary Tumorboard and in consideration of this benign finding, the oncological follow-up plan did not need to be adapted.

Discussion

Clinically, Littoral Cell Angioma can be associated with abdominal pain, splenomegaly, symptoms of hypersplenism and fever of unknown origin [4]. In our case, no other clue than the computed tomography findings suggested a splenic pathology. Radiologically, the features of littoral cell angioma have been well described, but the specificity is poor and it is not always possible to differentiate from other vascular neoplasms, metastases, disseminated infections or sarcoidosis [9]. Typically there is a splenomegaly, the lesions are multiple, hypodense in the native computed tomography scan relatively to the normal spleen parenchyma and with some enhancement during portal venous phase (what corresponds to blood-filled nodules) [10]. A histologic analysis is needed for the definitive diagnosis, which was

performed after the splenectomy in our patient. It is also possible to differentiate the littoral cell angioma from a hemangioma, a lymphangioma or a hamartoma preoperatively on the cytology of material from a fine-needle aspiration. This is however challenging due to the rare cytological descriptions [11]. The dual expression of endothelial and histiocytic markers helps differentiate it from the other vascular tumors (CD68+ / CD31+ / Lysosyme+ / vWF+ / CD21+ / CD34- / CD8-) [11]. The microscopic histopathologic analysis of our specimen showed anastomotic vascular channels with flat and bulged endotheliocytes with the typical immunohistochemical signature.

In the case of our patient, the littoral cell angioma was associated with a rectal cancer. A paper published in 2016 [9] confirmed the frequent association of the splenic neoplasia with other malignancies, such as colorectal carcinoma, renal cell carcinoma, endometrioid endometrial adenocarcinoma, chronic lymphocytic leukaemia, malignant lymphoma, pancreatic adenocarcinoma, multiple myeloma, prostate cancer, GIST of the stomach, carcinoma of the thyroid gland, breast cancer and melanoma. It also seems to be associated with many syndromes of immune dysregulation or systemic diseases known to cause immune disturbances [9]. The exact correlation between those disorders still remains object of further investigation [12]. Special to our case is the close radiological documentation of the growth of the Angioma over four years in the context of the follow-up of the rectal cancer, since an abdominal imagery was performed every 6 months. It showed a linear growth, with a diameter gain of about 4 mm per year (Graph 1).



Graph 1: Growth of the Angioma between April 2018 and December 2021. Abscissa: date. Ordinate: diameter of the main tumor in mm.

Given the incidental discovery of the tumor, we do not know how old it was at the time of the initial diagnosis, and if the growth is indeed always linear, or if the lesion grows faster when it is young or after a couple of years of development. An earlier and longer documentation would allow to answer this question, but since the Littoral Cell Angioma is an asymptomatic lesion, it will often not be diagnosed. Moreover, a longer documentation wouldn't be performed if it wasn't necessary for the follow-up of

a concurrent malignant condition, like in our case.

Conclusion

Littoral Cell Angioma should be considered as a differential diagnosis of red pulp splenic tumors, particularly if the lesions are multiple and if there is a splenomegaly. It is a very rare condition, but its prevalence is probably underestimated due to its asymptomatic course. The entity still needs to be better described cytologically. That then would allow diagnosing it by biopsy without mandatory having to perform a splenectomy given the benign nature of these tumors. A new study published in February 2022 suggested that an image-guided biopsy with follow-up be the optimal diagnostic choice for patients with Littoral Cell Angioma, but a pathological uncertainty is in the cytology always possible, and standard recommendations still need to be made for the diagnostic procedure [8].

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