Case Report

Angiolipomatous mesenchymal hamartoma (angiolipomatosis) of the sigmoid mesocolon

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Abstract: Background: Primary mesenteric tumors are exceedingly rare and may thus pose a diagnostic challenge. They encompass both benign and malignant neoplasms as well as reactive and idiopathic tumefactive fibroinflammatory lesions. Method and results: A 70-year-old man who was diagnosed with sigmoid colon cancer was found to have a large non-homogeneous predominantly fatty retroperitoneal soft tissue mass on computerized tomography (CT) scan. The mass was attached to the aorta and have encased the inferior mesenteric artery and extended into the sigmoid mesocolon. Histological examination of the mass showed ill-defined lipoma-like mature fatty tissue traversed by paucicellular fibrous septa entrapping small nerves and containing remarkably increased venous blood vessels reminiscent of soft tissue angiomatosis without evidence of malignancy. The histological features were consistent with an angiolipomatous hamartomatous mesenchymal proliferation. Conclusions: Angiolipomatous hamartoma might be histogenetically related to soft tissue angiomatosis. To our knowledge, this case represents the first well documented lesion of this type at this location and must be distinguished from other fat-containing masses, particularly angiomylipoma, sclerosing mesenteritis and mesenteric liposarcoma.

Keywords: Angiolipomatous hamartoma, mesocolon, angiomatosis, tumor-like lesion

Introduction

Primary tumors of the mesentery and the retroperitoneum are relatively uncommon and may thus pose a great diagnostic challenge [1]. They encompass a heterogeneous group of benign and malignant neoplasms as well as reactive and idiopathic mass-forming fibroinflammatory conditions [2]. As liposarcoma represents the commonest retroperitoneal sarcoma, radiological findings of a fat-containing retroperitoneal or mesenteric mass commonly raises concern about liposarcoma. This is particularly true for large mass lesions displaying both lipogenic and non-lipogenic components [3-5]. Based on the fact that radical surgical excision is the first and most effective therapeutic option for patients with retroperitoneal liposarcoma, it is necessary to distinguish a variety of benign fat-containing mass lesions that may closely mimic liposarcoma on imaging investigations. In this report, we describe the radiological and pathological features of a large hamartomatous lipofibrovascular mesenchymal proliferation detected incidentally on preoperative computerized tomography (CT) scan performed for staging of a newly diagnosed colorectal cancer and thought to be suspicious for liposarcoma.

Case history

A 70-year-old Caucasian man presented with signs of lower gastrointestinal bleeding. His past medical history was remarkable for alcoholic liver cirrhosis, cardiac arrhythmia with atrial fibrillation, Parkinson disease, diabetes mellitus, hypercholesterinaemia, status post aortic valve prosthesis 14 years ago and hyperthyroidism. Colonoscopy revealed an ulcerated tumor in the sigmoid colon consistent with colorectal cancer. The abdominal CT scan showed a large ill-defined non-homogeneous mass in the sigmoid colon mesentery encasing the inferior mesenteric artery and attached to the aorta.
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(Figure 1). The mass was predominantly fatty in nature with more centrally located non-lipogenic soft tissue component (Figure 1). To better judge the necessity for a more radical surgery, an intra-operative surgical biopsy was submitted for frozen section examination and revealed no evidence of malignancy. Accordingly the mass has been dissected and separated from the aorta thereby preserving the hypogastric plexus located ventral to the abdominal aorta. This was followed by mesocolic excision of the sigmoid and the descending colon together with partial mesorectal excision down to the upper rectum (Figure 2). The patient is alive and well with no evidence of recurrence of the mass or the colorectal cancer at last follow-up two years after surgery.

Results

The resection specimen was fixed in 4% buffered formalin and embedded routinely in paraffin for microscopic examination. Histologically, the sigmoid colon tumor showed a moderately differentiated adenocarcinoma (pT3b, pN0, L0, V0, R0, UICC stage IIA). The sigmoid mesentery showed an ill-defined tumor-like swelling of fatty tissue covered by glistening serosa (Figure 2). Histologically, the mass was composed of mature fatty tissue that contained abnormal aggregates of variably sized predominantly venous muscular blood vessels. These formed either confluent solid areas (Figure 3A) or were arranged into small lobules of capillary-sized muscular veins surrounded by a larger feeding vessel and scattered small nerve fibers (Figure 3B, C). Rare larger arterial vessels showed fibromuscular dysplasia with prominent intimal thickening. The remainder of the mass showed a predominance of fatty tissue traversed by paucicellular fibrous septa (Figure 3D). Small lymphoid aggregates, occasionally forming small follicles, and scattered few mononuclear cells were seen. The mass blended imperceptibly with the surrounding mesenteric fat. There were no areas of necrosis, prominent sclerosis, atypical or multinucleated stromal cells, lipoblasts, florid diffuse inflammation, plasma cells or foamy histiocytes. Epithelioid myoid cells as seen in renal-type angiomyolipoma were absent. Immunohistochemistry was completely negative for MDM2 or HMB45. Smooth muscle actin and desmin highlighted the muscular vessels, but there was no true muscular component within the fatty tissue.

Discussion

The histological features of the current lesion, particularly the prominent aberrant vascular channels in a background of abundant fatty tissue that gradually merged with the surrounding mesenteric and retroperitoneal fat and entrapped small nerve fibers are consistent with a benign hamartomatous etiology of the lesion and argue against a true neoplasm. Hamartomatous lesions of the mesentery are generally asymptomatic. Most are discovered incidentally

Figure 1. Abdominal CT scan in an axial view after intravenous contrast agents. Predominantly fatty (open arrows) mesenteric tumor with soft-tissue parts (white arrow) surrounding the inferior mesenteric artery (black arrow).

Figure 2. The resection specimen of the sigmoid colon cancer showed a large smooth-surfaced mass within the mesocolon (arrows) after preparation from the aorta (interrupted line).
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Figure 3. A. Histologically, the mass was predominated by crowded small-sized venous vessels in fibro-fatty background. B. In areas with predominant fat, vessels formed minute lobules. C. Higher magnification of a venous lobule showing a larger feeding vein on the left. D. Poorly vascularized fat lobules were separated by fibrous septa containing small veins.

on imaging procedures or during surgery for unrelated diseases and may pose a diagnostic challenge.

The differential diagnosis of mesenteric masses depends, among other clinicopathological features, particularly on the age of the patient and the imaging characteristics of the mass. Thus, omental-mesenteric myxoid hamartoma [6] is a possibility in children but not in adults and the elderly patients. Angiolipoma is an important consideration in our case. However, unlike the usually small and encapsulated angiolipoma and its prominent component of thin capillaries with occasional fibrin microthrombi, the lesion in our case was large, poorly circumscribed and merged with adjacent retroperitoneal fat and the vascular component was dominated by prominent muscular veins. A well differentiated liposarcoma was excluded based on lack of lipoblasts, nuclear atypia, atypical and bizarre stromal cells and absent MDM2 expression by immunohistochemistry [7]. The presence of a prominent aberrant vascular component in an intra-abdominal fatty mass raises the possibility of renal-type angiomylipoma. However, the normal-looking non-tortuous vessels, the absence of a clear-cut myoid component and the complete lack of reactivity for HMB45 and smooth muscle markers excluded a fat-predominant angiomylipoma [8]. Further differential diagnoses that were also excluded by appropriate histological criteria include fibrosclerosis mesenteric conditions [9,10] and mesenteric lipoma/lipomatosis [11]. The lesion in our case carries a superficial similarity to a recently described pediatric soft tissue tumor composed of an admixture of adipose tissue
and fibroblastic elements (hence referred to as lipofibromatosis) [12]. However, these lesions occurred almost exclusively in the peripheral soft tissue of children as superficial cutaneous or subcutaneous lesions and usually contain a clear-cut spindle cell fibrous component but lack a prominent vasculature. Thus the age of our patient and the histological features exclude this possibility.

The histogenetic classification of the current lesion raises several other considerations. An arteriovenous malformation is one possibility. Manger et al reported a case of an extensive venous angiomatous hamartoma of the mesentery that was complicated by chylothorax and chyloous ascites [13]. However, the mass was predominantly formed by abnormal tortuous blood vessels and fibrous tissue and a prominent fatty component was not present in their case. More importantly, the lobular pattern of the vascular channels in our case was not described in their case and fibrointimal thickening as seen in arteriovenous shunting was not a prominent feature in our case indicating that the two lesions are distinct. In our opinion, the current case showed histological features that are very reminiscent of the lesions reported by Rao and Weiss as soft tissue angiomatosis [14]. In that study (n=51 lesions), most lesions originated in the peripheral soft tissue and/or bone; only a single case has involved the retroperitoneum. The radiological features suggested a sarcoma in the cases with imaging findings. Histologically, the main features of soft tissue angiomatosis were a haphazard proliferation of thick-walled venous and capillary-sized vessels and absence of vascular wall hypertrophy/intimal thickening and thrombosis as would be expected in arterial shunting. Similar features were seen in our case. Furthermore, the presence of a prominent fatty component in several cases and of glomus cell proliferations in one case in that series pointed to a generalized mesenchymal proliferation. Thus, it is likely that some lesions in the group of soft tissue angiomatosis represent a hamartomatous proliferation (vascular mesenchymal hamartoma). Similarly, our case showed entrapment of small nerves suggesting a hamartomatous lesion. However, the older age of our case contrasts with the age range of soft tissue angiomatosis (more than 50% presented within the first two decades of life; the oldest patient was 54 years) [14]. Recurrence rate for soft tissue angiomatosis was 90% at a median interval of 5 years [14]. This is probably because most lesions have been partially excised. Up to now (two years from surgery), our patient has no evidence of recurrence.

In summary, we described a rare and unusual angiolipomatous (probably hamartomatous) tumor-like lesion in the sigmoid colon mesentery that posed diagnostic difficulty and raised pre-operative and intra-operative concerns about malignancy in an elderly patient with colorectal cancer. The biological potential and the histogenesis of this rare lesion remain to be further elucidated.

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