Case Report

Undifferentiated Pleomorphic Sarcoma Presenting as Abdominal Pain with a Pulsatile Mass

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Malignant fibrous histiocytoma (MFH) is a rare tumor that mostly involves adults aged 50 to 70. The most common anatomic location is the lower extremities. MFH of the retroperitoneum usually manifests late in its course and may be initially mistaken with other more common diagnosis. Here, the authors describe a 60-year-old man that was brought to the emergency department with a chief complaint of periumbilical abdominal pain. Our patient presented with symptoms consistent with a symptomatic aortic aneurysm, but a mass was encountered during surgery. In such circumstances the diagnosis of malignant sarcoma must be kept in mind and attempts at full resection with tumor-free margins are necessary.

1. Background

Malignant fibrous histiocytoma (MFH) was first described in 1961 by Kauffman and Stout as a histiocytic tumor with storiform growth in children [1]. Its malignant nature combined with an uncertainty regarding its origin and ability to involve both bone and soft tissue has given it the characteristics of a mysterious tumor, which is now accepted as a formal diagnosis of "undifferentiated pleomorphic sarcoma not otherwise specified" [2]. Various subtypes of MFH have been recognized including storiform-pleomorphic, myxoid, giant cell, and inflammatory [3]. Of these the storiform-pleomorphic subtype is the most common accounting for around 70% of the cases.

This rare tumor mostly involves adults aged 50 to 70, with a slight tendency towards male gender. The most common anatomic location is the lower extremities and the most common presentation is a growing painless soft tissue lump. Here we present the case of a 60-year-old man who came to the emergency department with a complaint of abdominal pain.

2. Case Report

A 60-year-old man was brought to the emergency department with a chief complaint of periumbilical abdominal pain. He suffered from abdominal pain for two weeks but it was significantly increased in the three days prior to admission. The pain radiated to the back and no accompanying signs or symptoms were present. It was described as a constant pain without any alleviating or exacerbating factors. The patient was a heavy smoker without any history of previous surgery or drug use.

The patient showed stable vital signs and a nuisance general appearance. No objective signs of weight loss or cachexia were observed. The physical exam was normal in the head and neck, chest, and extremities. In the abdomen a pulsatile mass was palpated in the periumbilical region. Routine laboratory tests revealed mild normocytic normochromic anemia, normal ESR and CRP, and liver function and diagnostic tests within normal limits.

Due to the pulsatile nature of the mass a CT angiography of the abdomen was ordered to look for vascular pathology. The tomography revealed a 5-by-4 cm pseudoaneurysm with...
hematoma formation in the infrarenal aorta alongside diffuse
dilation of the infrarenal inferior vena cava with extension
into both common iliac veins (Figure 1), and the patient was
prepped for surgical repair.

During surgery a pulsatile mass 20 by 20 cm in diameter
with surrounding hematoma was observed in left zones 1 and
2. Upon further investigation a huge retroperitoneal mass 11
by 8 by 6 cm in diameter with apparent local invasion to the
aorta and the inferior vena cava was found. En bloc resection
was attempted along with involved vascular segments. The
abdominal aorta was reconstructed using 16 mm-by-8 mm
Dacron bifurcated graft, while the IVC was ligated below the
renal veins but perhaps because of chronic process of disease
no lower extremity edema happened and the patient was
discharged after a week.

Pathologic evaluation of the specimen showed an undiff-
ferentiated pleomorphic sarcoma (a.k.a. malignant fibrous
histiocytoma) with a histologic grade according to French
Federation of Cancer Centers Sarcoma Group III (tumor
differentiation: III, mitotic rate >20/HPF: III, and tumor
necrosis <50%: I) (Figure 2). There was no evidence of lymph-
vascular or perineural invasion. Aneurysmal wall resection of
the abdominal aorta with blood clot and atherosclerosis was
also reported. The specimen was also positive for vimentin
(patchy areas), SMA (small portions), and CD68 (most parts).

3. Discussion

Malignant fibrous histiocytoma is a sarcoma of mesenchymal
origin affecting soft tissues of the body, particularly the
extremities and retroperitoneum, yet it has been reported in
almost all parts of the body [4–7]. The term malignant fibrous
histiocytoma is believed to be a misnomer since the precise
origin of MFH cells has been disputed and the concept of
fibrohistiocytic differentiation has been challenged [8]. For
this, the WHO has defined the tumor and its subtypes under
undifferentiated pleomorphic sarcoma (UPS) not otherwise
specified (NOS). Most undifferentiated high-grade pleomor-
phic sarcomas and undifferentiated pleomorphic sarcomas
with giant cells occur in the deep soft tissues of the extremities
while undifferentiated pleomorphic sarcoma with prominent
inflammation is most commonly seen in the retroperitoneum
[2].

Retroperitoneal and intra-abdominal tumors may
present with constitutional symptoms, including fever,
malaise, and weight loss [9, 10]. The tumor is often large at
presentation since it usually goes unnoticed for a long time
and may cause displacement of the bowel, kidney, ureter,
and/or bladder. In our patient the tumor had displaced an
abdominal aortic aneurysm anteriorly thus creating the
unique clinical presentation of abdominal pain and pulsatile
mass.

Although the best imaging modality for evaluation of
the tumor is Magnetic Resonance Imaging (MRI), for intra-
abdominal masses usually this is not the initial step. Gener-
ally, retroperitoneal undifferentiated pleomorphic sarcomas
manifest as heterogeneous masses with areas of hemorrhage
and necrosis and occasionally focal or diffuse coarse calcifica-
tions. While invasion of the abdominal musculature is a well-
recognized phenomenon, the renal veins, or inferior vena
cava, are not invaded [11, 12]. In our patient too despite the
initial impression of vascular invasion in the operating room,
pathologic reports confirmed that the vascular invasion had
not occurred.

One must bear in mind that the pathological diagnosis
of MFH/UPS is a diagnosis of exclusion. In order to rule
out a pleomorphic nonmesenchymal neoplasm resembling
a pleomorphic sarcoma, pleomorphic sarcoma as a result
of dedifferentiation, and pleomorphic sarcoma with a spec-
cific line of differentiation, a combination of sampling and
immunohistochemistry should be used.

Microscopically, UPS has a highly variable morphologic
pattern and shows storiform to haphazardly arranged pleomor-
phic areas. In immunohistochemistry they show features
of fibroblasts/myofibroblasts [8].

The current treatment of choice for primary MFH/UPS
is wide surgical resection aiming at tumor-free margins.
While adjuvant radiotherapy has become an integral part of
treatment, the role of chemotherapy is still controversial in
these tumors [13–15].

4. Conclusion

MFH/UPS of the retroperitoneum usually manifests late in
its course and may be initially mistaken with other more
common diagnoses. Our patient presented with symptoms consistent with a symptomatic aortic aneurysm, but a mass was encountered during surgery. In such circumstances the diagnosis of malignant sarcoma must be kept in mind and attempts at full resection with tumor-free margins are necessary.

**Competing Interests**

The authors declare that they have no competing interests.

**References**


