A Case Report of an Intraabdominal Microcystic and Pseudopapillary Spindle and Round Cell Neoplasm and a Comparison to Other Intraabdominal Mesenchymal Tumors

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Abstract. Aim: We present a case of a new mesenchymal tumor entity named ‘distinctive microcystic and pseudopapillary spindle and round cell neoplasm’, of which only 30 cases have been reported worldwide. Case Report: A fifty-two-year-old woman presented in January 2012 with epigastric pain of changing character and weight loss. Examinations revealed a tumor 10×6.8×9.8 cm in diameter showing infiltration in the surrounding organs. A Whipple procedure and a right hemicolectomy were necessary to achieve free resection margins. At the last follow-up in November 2012 the patient was well and there were no signs of recurrence or metastatic spread. Intra-abdominal mesenchymal tumors cause unspecific abdominal symptoms. Local recurrence and metastatic spread can occur. The main prognostic factors for survival are a free margin after resection and the histological subtype of the tumor. Behavioral prediction is not possible in every case and the treatment has to be individualized for every patient. Conclusion: This case represents a new entity of mesenchymal tumor. It was treated according to the guidelines for intra-abdominal sarcomas. Further investigation of this kind of tumor is necessary to define therapeutic guidelines.

Intra-abdominal mesenchymal tumors are an inhomogeneous group of neoplasms originating from primitive mesenchymal cells. Many histological subtypes have been described, although they are relatively rare. These tumors are, therefore, a diagnostic challenge (1).

Mesenchymal tumors may recur locally and might metastasize. Complete surgical resection is the only curative treatment but may be limited by the involvement of adjacent structures (2). Only limited results for the use of chemotherapy and postoperative radiation regarding mesenchymal tumors exist. New multimodal treatments are also being used with varying success (3, 4). The main prognostic factor for survival, however, remains the margin of resection after surgery (2, 5). Behavioral prediction is still not possible in every case and the treatment has to be individualized for every patient.

The most common intra-abdominal mesenchymal tumors are the retroperitoneal liposarcoma and the gastrointestinal stromal tumor (GIST). GIST is the most common mesenchymal tumor of the gastrointestinal- tract (1, 6, 7).

Recently, new entities of mesenchymal tumors have been described in the literature. Examples for such tumors are desmoplastic small round cell tumors (DSRCT) and perivascular epitheloid cell tumors (PEComa) (8, 9).

Here, we present another new tumor entity. In the presented case, our patient was suffering from a distinctive microcystic and pseudopapillary spindle and round cell neoplasm, a tumor entity of which only about 30 cases have been described to date. Our patient was treated with complete surgical resection at our department.

Case Report

In January 2012 a fifty-two-year-old women presented with epigastric pain of changing character which had lasted for several weeks as well as weight loss. Clinical examination showed a tenderness of the upper abdomen without any palpable mass.

Abdominal sonography revealed an abdominal tumor most likely originating from the ventricular antrum and corpus of varying density values and solid portions.
The laboratory values were unremarkable, without elevation of any tumor markers. A gastroscopy was performed which showed a huge protrusion at the small curvature but a normal mucosal surface.

An explorative laparotomy was performed at another hospital. Due to the complexity of the tumor, only an open biopsy was carried out. The patient was then sent to our department for further treatment. The histological findings of the biopsy revealed a rare tumor entity named “distinctive microcystic pseudopapillary spindle and round cell neoplasm” (see Pathological Findings).

Magnetic resonance tomography (MRT) as well as magnetic resonance angiography and a magnetic resonance cholangiopancreaticography (MRCP) were performed and revealed a 10×6.8×9.8 cm hyperintense tumor in contact with the stomach, pancreas, liver hilum and transverse colon (Figure 1). The tumor was also in contact with the superior mesenteric artery, the gastroduodenal artery and the proper hepatic artery, without any signs of infiltration.

After the patient had given informed consent, transverse upper-abdomen laparotomy was performed. The intraoperative site showed an infiltration of the tumor into the transverse mesocolon, as well as the stomach and the pancreas (Figure 2). A Whipple gastropancreaticoduodenectomy and a right hemicolectomy were performed without any intraoperative complications. The histological workup of the removed tumor was consistent with the biopsy obtained earlier. The resection margins were clear at all sites.

The postoperative course was without any severe complications. A clinical control examination as well as a computed tomography (CT) were scheduled three months after discharge.

The CT was performed in August 2012 and showed no signs of local recurrence or metastatic spread. The last follow up was in November 2012. The patient was well at that time and the recent CT didn’t reveal any changes to the previous examination.

Pathological findings. The macroscopic overview was of a large, soft tissue tumor mass infiltrating the pancreas. The cut surface revealed the white, fascicular, partly microcystic and myxoid structure of the tumor (Figure 3). Hematoxylin and eosin (HE) stain revealed the distinctive microcystic pseudopapillary spindle and round cell neoplasm, composed of mesenchymal, spindle-shaped cells with a solid and microcystic growth pattern of the tumor cells. The cell nuclei were round to oval with pale eosinophilic to dark basophilic surrounding cytoplasm. The tumor was highly vascularized, as demonstrated by a dense network of vessels lined by cluster of differentiation(CD)34-positive endothelial cells or composed of smooth muscle antibody (SMA)-positive smooth muscle cells. The proliferation rate, evaluated by MIB-1 proliferation index was moderate to focally high (Figure 4).

Discussion

Very little is known about the clinical behavior and management of an entity termed ’distinctive microcystic and pseudopapillary spindle and round cell neoplasm’. The tumor found in our patient was a large mass invading parts of the stomach wall, the duodenum and pancreas and the colon. Extensive en-bloc resection was therefore necessary. This growth pattern resembles that of a liposarcoma which is most often localized in the retroperitoneum and tends to infiltrate contiguous organs such as the small intestine, colon, kidney, adrenal gland or parts of the vena cava (10). Other mesenchymal tumor entities such as the DSRCT, which occurs mostly in young, male adults and PEComa show similar behavior (9, 11, 12). DSRCT develops in the abdominal cavity and invades the omentum with multiple peritoneal implants, whereas PEComa occurs most commonly in the retroperitoneum and

Figure 1. Magnetic resonance imaging image of the tumor showing a 10×6.8×9.8 cm hyperintense tumor in contact with the stomach, pancreas, liver hilum and transverse colon.

Figure 2. Intraoperative site showing the tumor.
at visceral sites, with a predilection for the gastrointestinal tract and the mesentery (9, 11).

Other than liposarcomas, GISTs do not often invade surrounding structures but rather tend to displace them and do not invade contiguous organs like in the presented case (2, 13).

The symptoms caused by the intra-abdominal microcystic and pseudopapillary spindle and round cell neoplasm were epigastric pain and weight loss, most likely caused by reduced oral intake. This is consistent with the symptoms caused by other abdominal mesenchymal tumors such as retroperitoneal liposarcomas. Symptoms are usually vague
and start to show after the tumor has reached a diameter of more than 10 cm (10, 13). The symptoms of GISTs largely depend on the location and the size of the tumor. Small tumors are often found incidentally during endoscopy. Larger tumors can cause nausea, pain, early satiety and can be the source of significant gastrointestinal bleeding (2).

The evaluation of our patient included physical examination, blood sampling including tumor markers, abdominal sonography and a gastroscopy. Furthermore in our case we used MRT-angiography to better-evaluate the relation between the tumor and the main vessel of the abdomen, which is very useful in evaluating the resectability of the tumor and to prepare the surgeon for difficulties arising from a possible infiltration of the tumor into the main vessels. An explorative laparotomy was also performed at another hospital to obtain the histological sample that was examined before the resection of the tumor. Because the resection was technically demanding, the patient was then transferred to our hospital.

The evaluation of patients with other mesenchymal tumors also includes physical examination, blood sampling including tumor markers, abdominal sonography and a gastroscopy if a GIST is suspected. MRT, however, is not routinely used. According to literature, fluorodeoxyglucose positron-emission tomography (FDG-PET) can be a helpful diagnostic tool. Because visceral sarcomas like GISTs are often submucosal, it is difficult to obtain biopsies endoscopically. Furthermore, these tumors are fragile and often hypervascular and there is a risk for bleeding. Therefore most GISTs are preoperatively diagnosed on clinical grounds (2, 13). Biopsies of retroperitoneal mesenchymal tumors should be taken CT-guided if necessary for confirmation of the diagnosis if resection is not possible or if the diagnosis would otherwise remain unclear (10).

In our case, surgical en-bloc resection was the treatment of choice and total resection of the tumor during the first operation offered the best chance for cure. Because of the infiltration of the tumor into the transverse mesocolon, as well as the stomach and the pancreas, a Whipple procedure as well as a right hemicolecotomy were performed.

Complete surgical resection is also the treatment of choice for other mesenchymal tumors. Since GISTs do not often invade surrounding structures but rather tend to displace them it is often possible to resect the tumor without removing adjacent organs. During resection, the pseudocapsule of the tumor should be left intact to prevent peritoneal dissemination. Furthermore, during the operation, the whole abdomen should be examined for metastatic spread, especially the liver and peritoneum, which are the most common sites for metastatic spread of GISTs. Any suspicious lesions should be resected because peritoneal metastases in particular are easily overlooked in preoperative imaging. Since lymph node metastases are very rare from GISTs, lymphadenectomy is not routinely indicated (2, 13). The therapy of liposarcomas which exhibit an infiltrative growth pattern similar to that of microcystic and pseudopapillary spindle and round cell neoplasm often make the resection of contiguous organs necessary (10).

There is no known adjuvant therapy for the described microcystic and pseudopapillary spindle and round cell neoplasm.

GISTs are successfully adjuvantly treated with the tyrosine kinase inhibitor imatinib which helps to control local recurrence and metastatic spread (2, 14). Oral intake of imatinib for 12 months after resection is advised for patients with high-risk GISTs and was shown to improve recurrence-free and overall survival (14). Imatinib is also used as neoadjuvant treatment in cases with primary unresectable GISTs and is the first-line of therapy if recurrence or metastatic spread occurs. The second-line agent is sunitinib, which is used if a GIST proves resistant to imatinib. New strategies in the therapy of GISTs also include the use of immunomodulation (14).

External-beam radiation after resection of retroperitoneal liposarcoma prolongs the timespan to local recurrence but does not seem to have an influence on patient survival. However, its use is difficult because of the great area which needs to be treated and complications arising from the radiation of sensitive areas such as the small intestine (10, 13). If a soft tissue sarcoma spreads throughout the abdomen, in the absence of any extra-abdominal dissemination, the term abdominal sarcomatosis applies (15). If clear resection margins cannot be achieved the best treatment option is intraoperative radiation during surgery. No radiation was used in our case because the risk of metastatic spread is very low and the resection margins were clear.

There is, so far, no evidence-based knowledge about the prognosis of intraabdominal microcystic and pseudopapillary spindle and round cell neoplasm. Out of the 30 cases which are known worldwide, only one patient had local recurrence and none showed signs of metastatic spread so far.

The prognosis of GISTs depends strongly on the mitotic rate, the size of the tumor and the location in the gastrointestinal tract. About 10 to 30% of all GISTs exhibit malignant behavior characterized by local recurrence or metastasis. The most common sites for metastasis of GISTs are the liver, the peritoneum and the lungs (1). Gold et al. developed a nomogram which takes into account mitotic rate, tumor size and location in the gastrointestinal tract and assesses the 2- and 5-year recurrence-free survival in patients undergoing potentially curative resection for primary GIST (16). The type of gene mutation found in the GIST can also influence the prognosis (14). Liposarcomas also have the potential for local recurrence as well as metastatic spread. The main prognostic factors for survival and recurrence after surgical resection concerning liposarcomas are the
histological subtype and the need for contiguous organ resection apart from the kidney (6). DSRCT exhibits local recurrence on a regular basis and complete surgical resection is often not possible. Three-year survival does not exceed 30-40% (11). PEComas can exhibit malignant behavior with lymphatic spread and the occurrence of distant metastases to the liver, lungs and bones (9).

**Conclusion**

“Distinctive microcystic and pseudopapillary spindle and round cell neoplasm” is a new entity of mesenchymal tumor. Since very little is known on the clinical behavior of this tumor, it was treated like a sarcoma. It is not known whether this kind of tumor is prone to spread systemically. With regard to its similarities to GIST and liposarcoma, it also tends to locally recur. Complete surgical resection with the resection of contiguous organs adherent to the tumor was the treatment of choice for our patient. There is no adjuvant therapy for this kind of tumor as yet. To detect local recurrence as early as possible, close follow-up intervals were set for our patient.

Further investigation of this new mesenchymal tumor entity is necessary to implement guidelines for treatment and to evaluate possible adjuvant therapies.

**References**