Mesenteric tumors: Diagnosis and treatment

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Summary Mesenteric tumors are rare and consist of a heterogeneous group of lesions. Masses may arise from any of the mesenteric components: peritoneum, lymphatic tissue, fat, and connective tissue. Cellular proliferation can also arise from infectious or inflammatory processes. They can be classified as solid or cystic, benign or malignant. Mesenteric tumors are usually discovered incidentally or during investigation of non-specific symptoms. While clinical examination and imagery may suffice to make the diagnosis, histopathology is often required by either needle percutaneous or surgical biopsy, or immediate excision. Therapeutic management options vary widely depending on the nature of the lesion; they range from simple observation or medical therapy to surgery. Benign well-delineated mesenteric masses that are symptomatic can often be treated by simple enucleation. But invasive malignant tumors require a carcinologic resection; a careful preoperative evaluation to assess the relationship between the mass and adjacent vascular and digestive structures is essential since they may dictate the need for extensive sacrifice of bowel with resultant intestinal insufficiency due to short bowel syndrome.

Introduction Mesenteric tumors are quite rare and include a diverse group of histologic entities with widely differing management strategies and prognosis. Their discovery is most often fortuitous or occurs during evaluation of vague non-specific abdominal symptoms. Computed tomography with IV contrast (CT) is the critical diagnostic imaging study. At times, the clinical context and laboratory findings may be sufficient to establish a definitive diagnosis but, in most cases, histopathologic diagnosis is necessary; this may require CT-guided needle biopsy, surgical biopsy, or immediate surgical resection. Management is dictated by the terrain, particularly the nature of the mass, its relation to adjacent anatomical structures, and associated symptoms.

Epidemiology Mesenteric masses are mostly of lymphatic origin. The incidence of cystic mesenteric masses is estimated at 1/100,000 in the United States, and half of these are cystic lymphangiomas [1]. Lymphoma is the most common solid mesenteric tumor [2]. Endocrine tumors of the small intestine are quite rare, but metastatic lymph node metastasis is present in 90% of these cases [3]. The annual incidence of desmoid tumors is 2.4–4.3/100,000 in the general population. The frequency of desmoid tumors in patients with familial polyposis ranges from 4–32% in various reports, but only 8% of desmoid tumors are localized to the mesentery [4].

The other types of mesenteric tumors are exceptionally rare with only sporadic case reports in the literature. Some lesions are more classically described in other locations and the mesenteric localization is exceptional (liposarcoma, inflammatory pseudotumor, Castleman’s disease, actinomycosis).

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The variety of mesenteric tumors

The wide diversity of different types of mesenteric masses is due to the multitude of cellular lines that compose the mesenteric structures: peritoneal surfaces, connective tissue, fatty tissue, lymphatic vessels, lymph nodes, and blood vessels. Each peritoneal leaflet is composed of a layer of mesothelial cells with villous protrusions and vesicular indentations. The subserosal tissue is rich in nerve endings, blood and lymphatic vessels, more particularly in the parietal surfaces than in the serosa overlying the viscera. Lymphatic vessels are very numerous and underlie up to 4% of the mesenteric peritoneal surface. Mesenteric masses arise either from a proliferation of the intrinsic cell lines (primary tumors) or from metastatic invasion (nodal metastases, carcinomatosis). They can also arise from cellular proliferation in response to an infectious or inflammatory process (actinomycosis, inflammatory pseudotumor).

Depending on their origin, one can distinguish the following types of mesenteric mass.

Tumors of lymphatic origin

Cystic lymphangioma (CL)

Cystic lymphangioma (CL) (Fig. 1) results from failure of lymphatic channels to develop drainage into the lymphatic system during embryogenesis resulting in lymphangiectasis and ultimately lymphatic cysts. Cyst content may be serous or chylous. Malignant transformation has been described in 3% of cases [5]. This is the most common mesenteric cystic tumor. Symptoms are often absent or minimal and non-specific; symptoms develop slowly but progressively with tumor enlargement. Symptoms may develop acutely (crampy abdominal pain, bowel obstruction) as a result of complications of CL: rupture, superinfection, intracystic hemorrhage, intestinal volvulus. CT imaging demonstrates a hypodense mesenteric cystic mass that may be unilocular, or multilocular with internal septations that may demonstrate contrast enhancement if they are thick. Magnetic resonance imaging (MRI) confirms the liquid nature of the contents and may demonstrate a fatty signal if the contents are chylous. A denser intracystic signal may be a sign of complications such as hemorrhage or superinfection.

Lymphoma

Lymphoma (Fig. 2) is the most common mesenteric malignancy [6]. In most cases, these are non-Hodgkins lymphomas [7] (40–67% are large B-cell tumors [8,9]). Systemic symptoms (fever, night sweats, weight loss) indicate advanced-stage disease. Complications such as intestinal obstruction, perforation and hemorrhage are rare [10]. Mesenteric lymphomas may remain asymptomatic for years, even when the tumor is voluminous. Staging studies should include examination of all peripheral lymph nodes and palpation to detect hepatic or splenic enlargement. The classical CT image of mesenteric lymphoma is a ‘sandwich’ appearance: a large agglomeration of homogeneously enhancing lymphadenopathy surrounding the mesenteric vessels with persistence of an intact perivascular fat border. The intestinal structures are displaced. At the periphery of the mass, venous stasis results in an infiltrated appearance of the mesentery. Calcification is not present unless the mass has undergone previous treatment. The presence of voluminous retroperitoneal lymphadenopathy argues strongly in favor of the diagnosis of lymphoma [11].

Figure 1.  CT with IV contrast: cystic lymphangioma—a multiloculated cystic mass with faintly enhancing walls.

Figure 2.  CT with IV contrast: lymphoma ‘sandwich’ appearance with extensive homogeneously enhancing lymphadenopathy enveloping the mesenteric vessels with preserved perivascular fatty envelope.

Castleman’s disease (CD)

Castleman’s disease (CD) is a benign lymphoproliferative disorder that only rarely involves the mesentery; its etiology is unknown. It is often seen in association with human immunodeficiency virus (HIV), and Herpes virus 8 (HHV8) infection, or in association with malignant pathologies such as Kaposi’s sarcoma, Hodgkin’s disease, and non-Hodgkin’s lymphoma. CD is usually localized (80%) and only rarely multifocal. Two subclassifications have been defined: a hyaline vascular type (the most common) and a plasmaocytoid type. This latter type results in systemic manifestations including general malaise, fever, anemia and elevated CRP levels. CT imagery demonstrates a mass with arterial phase enhancement and fine central microcalcifications. The principal tumor may be surrounded by satellite nodules [12].
Mesenteric nodal metastasis may present with local aggressivity with invasion of adjacent organs, resulting in the risk of local recurrence after simple surgical resection has been reported to be as high as 50%, and malignant degeneration was observed in one case after 10 years of progression [16].

Malignant peritoneal mesothelioma
Malignant peritoneal mesothelioma is a rare primary tumor. Only 6–10% of all mesotheliomas arise in the peritoneum, far exceeded in incidence by pleural and pericardial lesions. They classically occur in male patients in their sixties who have had extensive exposure to asbestos (60% of cases). Mesothelioma is a diffuse or disseminated malignancy, only rarely presenting as a localized tumor. An invasive mass which may have a cystic component; prognosis is somewhat better for this form of the disease. CT imaging shows a heterogeneous solid tissue mass, occasionally with a cystic component; uptake of IV contrast is inhomogeneous, and there may be invasion of adjacent organs but without diffuse peritoneal disease. The presence of calcification is much more uncommon than in pleural mesothelioma, however the association of calcified pleural plaques may help to indicate the correct diagnosis [17].

Tumors originating from fatty and connective tissue

Neoplasms
Solitary fibrous peritoneal tumors
Solitary fibrous peritoneal tumors (Fig. 4) are rare tumors of sub-mesothelial origin. They arise from the pleura in acellular fibrous tissue with no muscular or lymphatic components [14,15].

Benign cystic mesothelioma
Benign cystic mesothelioma is a rare tumor that most commonly affects young women. It is unrelated to asbestos exposure but may be associated with previous abdominal surgery or inflammatory diseases. It is typically located in the pelvis but mesenteric localization is possible. CT demonstrates a mass composed of multiple thin-walled cysts of variable size. Signs of local aggressivity with invasion of adjacent organs may be present. The term "benign cystic mesothelioma" is controversial since the behavior of these lesions over time is uncertain: the risk of local recurrence after simple surgical resection has been reported to be as high as 50%, and malignant degeneration was observed in one case after 10 years of progression [16].

Primary peritoneal tumors
Simple mesenteric cyst
Simple mesenteric cyst results from a congenital defect in the fusion of the peritoneal leaflets causing cystic formation in the mesentery. This rare entity is often diagnosed in childhood or adolescence. Cysts are typically small to moderate size (1–5 cm) and are asymptomatic; larger cysts may result in symptoms due to mass effect or complications. CT shows a simple unilocular cyst with no discernable wall thickness. Histologically, the cyst has serous content and the internal wall consists of mesothelial cells overlying

Metastatic lymphadenopathy from intestinal neuroendocrine tumor
The small intestine is the most frequent site of endocrine digestive tumors (Fig. 3). In 40–80% of cases, mesenteric nodal metastasis is present at the time of diagnosis [13]. These tumors secrete serotonin and growth factors that may result in the carcinoid syndrome (flushing, diarrhea) when hepatic metastases are present, and also in an intense desmoplastic reaction resulting in mesenteric scarring and retraction. Retractile mesenteric fibrosis with vascular encasement may result in bowel obstruction, segmental portal hypertension, and chronic mesenteric ischemia. CT with IV contrast demonstrates a poorly-delimited infiltrative mesenteric mass with associated calcifications and fibrous adhesive bands with outward radiating peritumoral vessels. The adjacent intestine may show signs of chronic ischemia and the absence of the perivascular "fat ring" helps to differentiate this from sclerosing mesenteritis. CT with enteric contrast may demonstrate localized intestinal wall thickening with early uptake of IV contrast. The presence of hypervascular hepatic metastases also helps orient the diagnosis toward an endocrine tumor. Metastatic lymphadenopathy may also be seen with small intestinal adenocarcinoma, but the clinical presentation is usually one of bowel obstruction caused by the primary tumor.

Figure 3. CT with IV contrast: small intestinal endocrine tumor (arrowhead) with retractile mesenteritis (arrow) corresponding to metastatic lymphadenopathy.

Figure 4. CT with IV contrast: solitary fibrous tumor of the peritoneum—well-delineated thick-walled tissue mass with hypervascular zones (arrows) and a cystic necrotic center.
two-thirds of cases and may be either benign or malignant. The mesentery is one of the extrathoracic locations of this tumor. CT shows a well-delimited soft tissue mass that may contain areas of necrosis or a cystic component. With IV contrast, there is marked heterogeneous enhancement during the portal phase [18]. Pathologic examination shows a mass of fusiform cells within a fibrous stroma overlaid by a layer of mesothelial cells. Characteristics of malignancy include hypercellularity, cellular atypia, an elevated mitotic index, tumor necrosis, and invasion at the margins. Solitary fibrous tumors in extrathoracic locations have a higher incidence of malignancy than those within the pleural cavity—between 11% and 55% in available reported series. Cranshaw et al. observed a recurrence rate after surgical resection exceeding 20% with a mean follow-up of 43 months, even when the tumors were not considered malignant at the time of initial resection [19]. These tumors have a potential for distant metastasis.

**Desmoid tumors**

Desmoid tumors (Fig. 5) are fibroblastic proliferations arising from fascial and aponeurotic tissue. They have a tendency to aggressive local invasion and to local recurrence but not to distant metastasis. They may occur sporadically, or in the context of familial adenomatous polyposis (FAP) or Gardner’s syndrome (FAP plus multiple osteomas, facial sebaceous cysts and desmoid tumors). These tumors are rare (0.03% of all tumors) [20]. Their annual incidence in the general population is estimated at three cases per million people per year [21]. They typically occur in the extremities, the neck, the thoracic wall, and the abdomen. Intra-abdominal desmoid tumors are even rarer (8% of all desmoid tumors) and typically involve the mesentery and small intestine. They are most commonly associated with FAP (70% of cases) and their incidence in this population ranges from 4–32% in various reports [22]. Factors favoring the development of abdominal desmoid tumors include local trauma, especially previous surgery. Thus, desmoid tumors arising in patients with FAP make their appearance after surgery in 75% of cases, usually within 3 years of the surgical procedure [20,23]. Hormonal factors also favor development and estrogen receptors have been identified in desmoid tumors. Genetic predisposition, as is evidenced by the association with FAP, underscores the need for genetic study of family members of any patient with a desmoid tumor. Desmoids may remain asymptomatic for prolonged periods and only be diagnosed incidentally by a radiologic study or laparotomy, or when enlargement eventually results in symptoms. CT imaging shows a well-circumscribed non-encapsulated soft tissue mass; it is of homogenous density before IV contrast injection, and only enhances slightly but often heterogeneously with IV contrast. MRI helps to assess the aggressiveness of the lesion; a hyperintense T2 signal is associated with hypercellularity [19,24]. Signs of poor prognosis include size (diameter > 10 cm), multiple tumors, infiltration or invasion of the mesentery, and encasement of the ureter [25]. Biopsy for pathologic diagnosis is controversial since it may result in accelerated tumor progression. Nevertheless, since management is often non-surgical, an accurate pathologic diagnosis is necessary. Abdominal localization of desmoid tumors is a sign of poor prognosis. Desmoid tumor is the second most common cause of death in patients with FAP (10%) [26,27].

**Fibromas**

Fibromas (Fig. 6) are well-delimited benign tumors that only rarely involve the mesentery. CT imaging shows a homogeneous well-delineated mass that may have a cystic component. There is enhancement of the periphery of the lesion after IV contrast injection. These tumors neither metastasize nor recur locally after complete resection with clear margins [28].

Tumors arising from mesenteric fat

**Benign lipomas**

Benign lipomas of the mesentery are very rare. Large lipomas may become symptomatic. Predisposing factors include obesity, hypercholesterolemia, diabetes, or a family history.
of lipomas [29]. Both CT with IV contrast and MRI can easily establish this diagnosis.

**Liposarcoma**

Liposarcoma (Fig. 7) is a common mesenchymal malignancy but mesenteric localization is rare. There are several histologic subtypes of various grade and gravity. The histologic classification use by the National Federation of Centers in the Fight against Cancer (FNCLCC) as described by Trojani distinguishes three grades of liposarcoma with direct relation to prognosis for global survival and to the incidence of distant metastasis [30]. These tumors are locally aggressive and have a high risk of local recurrence after apparently complete resection; the undifferentiated subtypes can be associated with distant metastasis. Characteristics on CT and MRI vary with the histologic subtype, generally show a non-homogeneous lesion with fat density intermixed with zones of denser tissue; the borders are poorly defined and often infiltrating. The density of the lesion exceeds that of subcutaneous fat. Contrast uptake is heterogeneous and progressive, varying with the subtype of liposarcoma [31]. Imaging is essential for staging. The degree of cellular differentiation and clear excision margins seem to be the major prognostic factors [32].

**Mesenteric stromal tumors: gastrointestinal stromal tumors (GIST)**

Mesenteric stromal tumors: gastrointestinal stromal tumors (GIST) (Fig. 8) are of mesenchymal origin, arising from the neuroendocrine cells of Cajal situated in the intestinal muscularis propria that act as the “intestinal pacemaker”. They arise most commonly in the stomach or small intestine, and more rarely in the esophagus, colon and rectum. Immunohistochemical staining demonstrates expression of CD-117 (95%) and CD-34 (70%) proteins; activating mutations of the C-kit and PDGFRA genes are frequently present. Extra-intestinal forms of GIST tumors are very rare (6.6% in a series of 142 GIST patients) [33]. GIST tumors have been found in the soft tissues of the omentum and mesentery, or more rarely in the pancreas, liver or gallbladder. Since the cells of Cajal are absent in these organs (except the pancreas [34]), this suggests that they develop from pluripotential precursor cells; their immunohistochemical and histopathological profile is identical to that of gastrointestinal stromal tumors (CD-117/CD-34 positive) [35]. Primary stromal tumors of the mesentery must be distinguished from peritoneal metastases from a primary GIST, which usually presents with multiple tumor nodules. CT imaging shows an enhancing mesenteric mass; the lesion is often heterogeneous due to intratumoral hemorrhage or necrosis, and may have a cystic component. These tumors are similar to GIST of the small intestine in terms of clinical progression and prognosis.

**Mesenteric cellular proliferations of infectious and inflammatory origin**

**Inflammatory pseudotumors or myofibroblastic tumors**

Inflammatory pseudotumors or myofibroblastic tumors are found almost uniquely in the lungs of infants. These are rare tumors and localization to the mesentery is exceptional. Originally considered to be an inflammatory reaction due to surgery or minor trauma, an occult infection, or an associated manifestation of systemic inflammatory disease, they are now classified as neoplasms since the discovery that the ALK gene is present in 50% of cases. This offers a better explanation for their tendency to local recurrence and their potential for invasion or distant metastasis. Slowly developing symptoms include abdominal pain, fever, malaise, weight loss, and anemia. CT imaging shows a well-delimited non-encapsulated soft tissue mass; there may be evidence of infiltration of the adjacent small bowel and mesentery. IV contrast uptake is quite variable. Central necrosis and calcification may be present as well as regional lymphadenopathy. While imaging alone may not be sufficient to make a definitive diagnosis, it also is useful for guiding needle biopsy of an unresectable mass [18]. Definitive diagnosis depends on histopathology, which demonstrates a proliferation of fusiform myofibroblasts mixed with inflammatory cells.

**Sclerosing mesenteritis**

Sclerosing mesenteritis is a rare benign inflammatory disease of unknown cause that can affect the mesentery and meso-colon; it occurs twice as frequently in men as in women.
It is subdivided into three successive stages based on its histologic evolution:

- mesenteric lipodystrophy corresponding to the first stage in which fat necrosis is the predominant lesion;
- mesenteric panniculitis corresponding to the second stage and is characterized by an intense inflammatory reaction (Fig. 9);
- retracted mesenteritis is the final stage in which fibrosis predominates.

Sclerosing mesenteritis may be associated with other pathologies such as retroperitoneal fibrosis or sclerosing cholangitis. It may also be seen in association with malignancies such as small intestinal neuroendocrine tumors, lymphomas, carcinomas of the breast, lung, colon or melanoma (in 69% of cases according to Daskalogiannaki et al. [36]). Its discovery is fortuitous in half the cases when abdominal imaging is performed for some other indication or it is diagnosed incidentally during abdominal surgery when a thickened retracted mesentery with nodularity is noted. The radiologic appearance varies with the stage of disease, i.e., fat necrosis, inflammatory change, or fibrosis. CT imaging shows a solid mass encasing the mesenteric vessels, occasionally with evidence of collateral circulation. Two important characteristics are the persistence of a fatty halo around the vessels (“fat ring” sign), and the presence in over 50% of cases of a pseudocapsule (better defined by MRI). Calcifications may be present, particularly in areas of tumor necrosis. Cystic degeneration has also been described. At times, imagery shows only a hyperdense appearance of the mesentery with small nodular lesions but no predominant tumor mass ("misty mesentery").

Actinomycosis

Actinomycosis is a chronic suppurative and infiltrating infection caused by Actinomyces israelii, an anaerobic Gram-positive bacteria. It results in a tumor mass made up of multiple abscesses with abundant granulation tissue that invades by direct extension across tissue planes; these characteristics make it difficult to differentiate from invasive neoplasms. A. israelii is part of the normal bacterial flora of the mouth, the intestinal tract and the female genital tract (particularly in the presence of an intrauterine contraceptive device [IUD]). The microorganism is not ordinarily pathogenic in immunocompetent patients, however, a mucosal breach due to trauma, foreign body, surgery, intestinal perforation, or inflammatory bowel disease may predispose to infection. The presence of an IUD is a risk factor [37]. Infections occur in cervicofacial (>50%), thoracic (20%) and abdominopelvic (20%) locations. Abdominal actinomycosis is usually related to perforated appendicitis or to small intestinal diverticulitis. While actinomycosis is undeniably a benign disease, it can complicate an underlying malignancy that served as the initial infection site. The clinical course is usually sub-acute with symptoms of abdominal pain, mass, fever and weight loss evolving over a period of days to months; an acute presentation of peritonitis may occasionally be seen related to a complication such as perforation or fistula. Laboratory findings suggest chronic inflammation: leukocytosis, elevated CRP, anemia, thrombocytosis. CT imaging reveals a poorly-delineated heterogeneous mesenteric mass that may be solid, cystic or mixed; there is intense enhancement in the solid portions with IV contrast; this may be associated with localized thickening of the intestinal wall. One particularity of this tumor is its ability to invade across anatomic planes with involvement of multiple compartments (abdominal wall, colon, retroperitoneum with ureteral compression). This unusual aspect of local invasive aggressivity in a setting of minimal symptoms and only mild signs of systemic disease should lead to consideration of the diagnosis of actinomycosis. Septic embolization may occur resulting in renal or hepatic abscesses. While a definitive diagnosis cannot be made on the basis of imaging alone, CT may help in guiding needle biopsy of the mass with evidence of actinomycotic "sulfur granules" on microscopic examination and culture of Actinomyces in the pus. However, the microorganism is fastidious and difficult to culture resulting in a 75% false negative result on bacterial culture. In reality, the diagnosis is not made until postoperatively in 90% of cases [38].

Whipple’s disease

Whipple’s disease is an infection caused by the Gram-positive bacillus Tropheryma whipplei. Several organ systems may be involved resulting in neurologic symptoms (myoclonus), arthritis, ophthalmologic symptoms of uveitis, and digestive symptoms (malabsorption) within an overall context of fatigue and malaise. Small intestinal involvement is associated with bulky mesenteric lymphadenopathy (Fig. 10). CT shows small intestinal wall thickening and associated clusters of enlarged lymph nodes with central hypodensity due to lipid infiltration. The diagnosis is confirmed by small bowel biopsy with demonstration of the microorganism by PCR [39,40].

Cystic lesions of extramesenteric origin

Mucinous cystic tumors

Mucinous cystic tumors develop from ovarian cells. Localization within the mesentery is unusual and may be explained by cellular implantation during embryologic ovarian migration, by mesenteric ovarian dysplasia, or by supernumerary ovaries. CT scan defines the location and relationship of the cyst to adjacent organs and may detect signs of malignancy: large size, multicocular tumor, thickened cystic wall, calcifications. Only examination of the resected lesion can
confirm the diagnosis and define tumor grade upon which all therapeutic decisions will be based [41].

Non-pancreatic pseudocysts

Non-pancreatic pseudocysts are cystic masses bounded by a fibrous non-epithelialized wall; they develop from a mesenteric hematoma or abscess that fails to reabsorb. Ultrasound or CT images show a thick-walled cystic mass with internal loculations containing debris or a bi-layer fluid level suggesting bloody or purulent sediment [15].

Dermoid cysts or mature cystic teratomas

Dermoid cysts or mature cystic teratomas are rare benign tumors that are usually diagnosed in childhood and that can occasionally localize in the mesentery. They arise from pluripotent cells and may contain a great variety of tissues (cartilage, bone, teeth, cutaneous tissue, fat...). CT findings are often pathognomonic [42].

Differential diagnosis

Except for peritoneal mesothelioma, which can be well-localized, primary and secondary peritoneal tumors are usually disseminated. Pseudomyxoma peritonei typically presents with ascites whose mucinous nature is suggested by non-homogeneous distribution through the abdominal cavity. Peritoneal carcinomatosis may develop synchronously or metachronously with the primary tumor, usually an intestinal or gynecologic carcinoma. The clinical history, symptoms related to the primary tumor, and the presence of ascites often suggest the diagnosis.

Intestinal duplications are another element in the differential diagnosis of cystic tumors of the mesentery. Classically, they come to light in childhood; they are usually in direct contact with the small intestine, but occasionally they may become detached and migrate into the mesentery. Ultrasound examination is often useful, demonstrating a sonoluent cystic mass with a multilayered wall similar to that of the adjacent intestine.

Management of a mesenteric mass

Even though mesenteric tumors are rare, the intestinal surgeon is frequently called upon to manage these lesions, both diagnostically and therapeutically. While the diagnosis is often made incidentally as the result of an imaging study, the most common symptom leading to diagnosis is abdominal pain or mass, with or without abnormalities of intestinal transit.

Diagnostic management

Analysis of imaging studies

While ultrasound is often performed initially, CT imaging remains the essential study. MRI is usually a complementary study unless there are contraindications to CT or IV contrast. MRI has the advantages of avoiding radiation exposure and offering high-resolution imagery. T1-weighted sequences in phase and phase-contrast, T2-weighted and gradient echo are particularly suited to highlight fat, fluid or hemorrhagic components.

![Figure 11. Radiologic diagnosis of mesenteric cystic tumors.](image-url)
The radiologist’s interpretation of these images should allow definition of:

- purely cystic masses: density analysis (serous, fatty, mucinous), the presence of internal septations, a solid tissue component, the pattern of enhancement after injection of IV contrast, and the delimitation of the tumor borders will permit the radiologist to propose a focused list of hypothetical diagnoses (Table 1, Fig. 11); because of the frequency of its occurrence, lymphoma usually heads this list;
- solid masses: the presence or absence of fatty content, the behavior of the tumor after injection of IV contrast, and the delineation of the tumor border allow the radiologist to propose a certain number of hypotheses: again, because of the frequency of its occurrence, lymphoma heads the list. Imaging characteristics allow the radiologist to suggest the diagnoses of mesenteric sclerosis, desmoid tumor or lipoma with high probability (Table 2);
- mixed solid-cystic masses: these often have a worrisome appearance suggestive of sarcoma or malignant lymphadenopathy with necrosis, but also of infectious or inflammatory pathologies (Table 2, Fig. 12).

Etiologic diagnosis

Clinical findings supply critical elements to assist in diagnosis and to orient therapeutic management. The simple discovery of an abnormal image during the diagnostic work-up of abdominal pain does not prove that the lesion is actually the cause of the observed symptoms, particularly in view of the frequency of other functional intestinal disorders. The clinician must precisely analyze the symptomatology.

In order of frequency, cystic lymphangioma is the most common diagnosis. Previous exposure to asbestos or the presence of calcific pleural plaques strongly orients diagnosis toward malignant peritoneal mesothelioma. A pseudocyst should always be considered in patients with a history of trauma, pancreatitis, or intra-abdominal abscess.

During the work-up of a solid mesenteric tumor, systemic symptoms such as fever and night sweats point to the diagnosis of lymphoma, which is the most common solid tumor, particularly in immunosuppressed patients. A past history of familial adenomatous polyposis or Gardner’s syndrome suggests desmoid tumor, especially in a patient with previous abdominal surgery. Lipomas occur more frequently in obese diabetic patients. Symptomatic carcinoid syndrome should suggest sclerosing mesenteritis related to a small intestinal carcinoid tumor. Radiologic evidence of aggressive invasion across tissue planes, in a patient in otherwise good general health except for a low-grade fever should suggest actinomycosis, particularly in a female with an IUD. The abdominal form of Whipple’s disease is associated with intestinal malabsorption and symptomatology involving other organ systems.

Laboratory data rarely contributes much to a specific diagnosis: but measures of systemic inflammation (CRP, WBC), an elevated LDH level, or elevated tumor markers (CA 125, CA 19-9) may be useful indicators.

Diagnostic approach

Sometimes a specific diagnosis can be confirmed by imagery; this is often the case for cystic tumors, sclerosing mesenteritis, or desmoid tumor in a patient with FAP. More often, multiple possibilities are evoked in the differential diagnosis and histologic evidence is required to establish the diagnosis. The different diagnostic modalities should be discussed in a multidisciplinary setting taking into consideration the list of possible diagnoses, whether the mass is symptomatic, whether it is resectable, and whether it is accessible to percutaneous biopsy. Needle aspiration or biopsy, often with radiologic guidance has a definite role in this setting; it is simple, rapid, and less invasive than surgical biopsy. Needle biopsy can be guided by either CT or ultrasound. Ultrasound guidance is less expensive, more rapid, and avoids the drawbacks of radiation exposure. It may also be safer since the needle can be constantly visualized in real time adjusting the needle trajectory to avoid vascular injury with the help of color Doppler imaging. Abdominal compression lessens the distance the needle must cross to reach the mass [43].

The choice between needle biopsy and needle aspiration depends on imaging characteristics of the tumor; either or both approaches may be used. Masses that are small, cystic, in close proximity to vascular structures, or appear to be abscessed should undergo needle aspiration. Larger solid masses should undergo needle core biopsy. The biopsy needle should be directed toward the zones of the tumor that enhance with IV contrast on CT or that are most echogenic on ultrasound examination. Heterogeneous areas may correspond to areas of tumor necrosis. Several biopsies can be performed along the same trajectory by insertion of an oversheath up to the edge of the mass through which the biopsy needle may be repeatedly passed. The usual contraindications to biopsy are abnormal hemostasis, gaseous distention, or lack of patient cooperation. Complications are rare (2—3%) [44] and usually well tolerated. There is a risk of intestinal perforation, but it is usually of little consequence due to the small caliber of the biopsy needle. Finally, the risk of tumor dissemination along the needle track is very low (<0.01%) [45].

The main objective of biopsy is to avoid misdiagnosis of a malignant tumor. While the specificity of needle biopsy to determine malignancy is excellent (100%), the sensitivity is considerably weaker (89—95%) [43,46]. Some authors propose that if a biopsied lesion is labelled as “benign” but without a specific etiologic diagnosis, the patient should undergo repeat biopsy [44]. When a difficulty-placed mass is inaccessible to percutaneous biopsy or if radioguided biopsy fails to yield a diagnosis (4% of cases according to Gottlieb et al. [47]), surgical biopsy by either laparoscopy or open laparotomy is the next option. This approach has the advantage of obtaining a larger tissue specimen, particularly for lymphomas where a large sample is necessary for tumor characterization.

Immediate frozen-section microscopic exam should ideally be performed to guarantee that the specimen is adequate for diagnosis; therapeutic decisions should be delayed, if possible, until a definitive diagnosis is established. Immunohistochemical and molecular PCR studies are useful to characterize certain histologic subtypes; unusual and rare entities with a risk of diagnostic error (neuroendocrine tumors, sarcomas, desmoid tumors) should undergo outside review by specialist pathologists.

The final objective is to select the best-adapted treatment for the particular diagnosis and to avoid unnecessary resection (lymphoma, actinomycosis, panniculitis) or potentially harmful surgery (desmoid tumor). For an asymptomatic tumor, the problem is to rule out a malignant or premalignant histopathology. For symptomatic tumors, the problem is to choose the most appropriate treatment for a given diagnosis, which may, in some cases, be non-surgical. Biopsy must be considered in the context of the tumor’s
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Table 1  Radiologic characteristics of mesenteric cystic masses.

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Tumors for which surgical resection is indicated

Lymphomas, for which chemotherapy is the primary treatment.

Lymphangiomas, when symptoms are minimal or complete resection is improbable, since the risk of local recurrence is 10−15%. Repeated intracystic injection of sclerosing agents may control symptoms when complete resection is impossible [49].

Sclerosing mesenteritis, in the early stage of fat necrosis, since there is a tendency to spontaneous resolution without treatment. While the progression of mesenteric panniculitis is more variable, it evolves favorably without treatment more often than not — over an interval of months to years, the inflammatory process tends to decrease returning eventually to normal. Medical treatment with immunosuppressive agents is indicated for severe cases with signs of systemic illness. When mesenteric panniculitis or the progressive fibrotic form of retractive mesenteritis results in bowel obstruction, surgery may be necessary (bypass, intestinal stoma). But even in the retractive mesenteritis stage, the clinical course is usually benign and stable [50].

Whipple’s disease is treated medically with trimethoprim-sulfa for a minimum period of 9 months [39].

Tumors for which surgical resection is debatable

Modern imaging techniques have resulted in the fortuitous discovery of very small lesions. Some authors recommend observation for small lipomas and fibromas until the tumor increases in size or becomes symptomatic. No size limit has been defined. This attitude is controversial in the era of minimally-invasive surgery, since resection spares the patient from the need for prolonged follow-up and the risk of eventual complications due to mass effect (volvulus) [51].

The management of desmoid tumors is still very debated. Asymptomatic desmoids should be observed initially, since 10% of these lesions will regress spontaneously. Symptomatic but uncomplicated tumors can be resected if they are small, well-circumscribed, and do not invade adjacent structures. For rapidly evolving or non-resectable desmoids, medical therapy with NSAIDs, hormones, or chemotherapy has been recommendable.
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used. As a last resort, subtotal surgical resection followed by radiation therapy of the residual tumor has been proposed [4]. Management algorithms have been proposed by Elias et al. for sporadic desmoids [52] and by Latchford et al. for desmoids associated with FAP [53].

The surgical procedure

The type of surgical procedure is determined by the nature of the mass and its extent. Simple enucleation is sufficient for well-circumscribed tumors. Infiltrating tumors require wider resection, which may be limited by the need to spare the blood vessels at the root of the mesentery, and by the amount of remaining small bowel. Short bowel syndrome (< 1.5 m) or ultrashort bowel (< 1 m) result in chronic intestinal insufficiency requiring medication therapy, optimization of oral nutrition, or even parenteral nutrition. The malabsorption syndrome is better tolerated if the ileocecal valve (which slows transit) and the right colon (which can adapt to an absorptive role, particularly of fats) can be preserved [54,55]. Other complementary options include segmental small bowel reversal to slow transit.

In certain circumstances, tumor debulking has been proposed. Thus, for small intestinal carcinoid with retractile mesenteritis where complete resection is impossible or for a non-resectable mesenteric mass, mesenteric debulking combined with resection of the primary tumor may be indicated in order to prevent ischemic or obstructive complications and to improve quality of life and survival, even if hepatic metastases are present [3,56]. For a non-resectable Castleman’s tumor, tumoral debulking is indicated followed by observation because of the risk of malignant transformation or by radiotherapy [57]. A preoperative diagnosis of actinomycosis permits the avoidance of aggressive resection, but a surgical procedure may still be useful for debridement of necrotic tissue, drainage of abscesses, and diverting ostomy in case of bowel

![Figure 12. Radiologic diagnosis of solid mesenteric tumors.](image)

![Figure 13. Role of percutaneous biopsy in the diagnosis of mesenteric tumors.](image)
obstruction. Decreasing the bacterial load permits a shorter duration of antibiotic treatment and decreases the risk of recurrence.

Conclusion

The management of mesenteric tumors is complex due to the wide variety of pathologic entities. Decision-making depends largely on analysis of imaging studies and should be addressed in a multidisciplinary setting where the surgeon plays a major role with regard to both diagnosis and treatment. The essential elements in choosing a therapeutic approach are the resectability of the mesenteric mass, the price to pay to achieve an R0 resection, and the degree to which the mass is symptomatic.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References

Mesenteric masses. Peritoneal neoplasms


