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REVIEW

Peritoneal lymphomatosis: CT and PET/CT findings and how to differentiate between carcinomatosis and sarcomatosis

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Abstract

Peritoneal lymphomatosis is a rare manifestation of lymphoma, seen most frequently with non-Hodgkin lymphoma, and it is important to be familiar with this condition, because early diagnosis directly affects the management of patients. This review illustrates the spectrum of imaging findings in peritoneal lymphomatosis, highlighting the use of positron emission tomography/computed tomography, showing common and uncommon subtypes of lymphoma associated with this entity, and how to differentiate it from peritoneal carcinomatosis and peritoneal sarcomatosis.

Keywords: *Peritoneal lymphomatosis; extranodal lymphoma; peritoneal sarcomatosis; peritoneal carcinomatosis; positron emission tomography.*

Introduction

Extranodal lymphoma refers to lymphomatous involvement in sites other than the lymph nodes, tonsils, thymus and Waldeyer lymphatic ring. It has an incidence of approximately 40% in lymphoma patients and can occur in any organ, however involvement of the peritoneal cavity is a rare clinical presentation. Extranodal lymphoma is a poor prognostic indicator overall[1,2]. According to the published data, the gastrointestinal tract is involved in 10–30% of patients with non-Hodgkin lymphoma (NHL) and involvement of the solid organs varies from 20 to 40% in the spleen to 4% in the adrenal glands[1]. An autopsy series of 322 patients with NHL aimed to demonstrate the incidence of omental involvement, and they found evidence of involvement in 64 patients (20%)[2,3].

Peritoneal lymphomatosis is defined as the intraperitoneal spread of lymphoma. Lymphoma does not usually involve the omentum, which is a peritoneal fold, because it consists of fibrofatty tissue devoid of lymphoid tissue[4]. The route of peritoneal dissemination is unclear, although in cases of intestinal lymphomas, spread is believed to be contiguous via the transverse mesocolon, the gastrocolic ligament and visceral peritoneal surfaces[2,3].

Most peritoneal neoplasms are malignant, and secondary malignancies far exceed primary tumors. The peritoneal surface may be secondarily affected by 3 cell lines: epithelial (carcinomatosis), mesenchymal (sarcomatosis) and lymphoid (lymphomatosis). Although these entities share some similar radiologic features, there are some contributory findings that may support the final diagnosis of lymphoma that the radiologist should know (Fig. 1). Peritoneal lymphomatosis is treated non-surgically, and often shows dramatic improvement with chemotherapy, and therefore its early and precise diagnosis is of utmost importance[2,3].

The purpose of this article is to review the computed tomography (CT) findings of peritoneal lymphomatosis, highlighting the use of [^{18}F]fluorodeoxyglucose (FDG)-positron emission tomography (PET)/CT, illustrating common and uncommon subtypes of lymphoma associated with this entity, and how to differentiate it from peritoneal carcinomatosis and peritoneal sarcomatosis.

	Peritoneal Lymphomatosis	Peritoneal Carcinomatosis	Peritoneal Sarcomatosis
Peritoneal lesions	Bulky homogeneous masses	Multiple small nodules	Bulky heterogeneous masses
Lymphadenopathy	Diffuse enlarged LNs	LNs around primary mass	None
Ascites	+	++	+/-
Examples			

LNs – lymph nodes

Figure 1 **Differences between peritoneal lymphomatosis, peritoneal carcinomatosis and peritoneal sarcomatosis.**

Histological subtypes associated with peritoneal lymphomatosis

Lymphomas are broadly subdivided into Hodgkin lymphoma and NHL, based on distinct clinical and histologic features. A detailed discussion of the histologic subtypes of lymphomas is beyond the scope of this article. Besides the pathologic classification, NHL subtypes are also grouped according to their clinical behavior as indolent, aggressive and very aggressive. In the long term, the longevity of patients with the highly aggressive and aggressive groups is better than that of the indolent group, because cure is almost not achievable in the latter[5]. Examples of indolent lymphomas are follicular lymphoma, small lymphocytic lymphoma, mantle cell lymphoma, lymphoplasmatic lymphoma, and some subtypes of marginal zone lymphomas. Aggressive lymphomas include diffuse large B-cell lymphoma, many natural killer cell lymphomas and most peripheral T-cell lymphomas. The entities included in the highly aggressive group are Burkitt lymphoma and lymphoblastic lymphoma. Follicular lymphoma and diffuse large B-cell lymphoma account for more than half of the cases of NHL. In the literature, peritoneal lymphomatosis is most commonly reported in association with diffuse large B-cell lymphoma[1–3]. Unlike NHL, Hodgkin lymphoma tends to spread by contiguous lymph node involvement instead of multifocal nodal involvement[6], and peritoneal lymphomatosis is an extremely rare manifestation of Hodgkin lymphoma.

CT imaging features

Omental caking is the most characteristic imaging presentation of secondary peritoneal malignancy. Omental caking may manifest as fine nodular, soft tissue studding or large confluent soft tissue masses within the omentum. The pattern most commonly found in peritoneal lymphomatosis is omental caking with bulky homogeneous masses (Fig. 2). Another characteristic imaging feature of peritoneal lymphomatosis is a homogeneous smooth thickening, diffusely infiltrating the peritoneum and the leaves of the mesentery (Fig. 3). Small omental nodules associated with fine infiltration of the omental fat producing a smudged appearance can also be encountered (Fig. 4). A stellate appearance of the mesentery is frequently seen and represents the results of an infiltrating process, causing thickening and rigidity of the mesentery (Fig. 5)[2]. Ascites can also be present. A retrospective evaluation of a small group of patients with peritoneal lymphomatosis found that most of the patients had mild to moderate ascites[3]. Additional findings may include peritoneal enhancement and peritoneal thickening, whether linear or nodular (Fig. 6). Enlarged lymph nodes are commonly seen in association with the mesenteric disease and should be a clue to the diagnosis. Isolated peritoneal involvement without accompanying bowel involvement or significant lymphadenopathy with lymphoma is unusual[7]. Associated lymphomatous involvement of the other intra-abdominal organs can also be present (Figs. 6–8).

Peritoneal lymphomatosis can be seen as an initial presentation of lymphoma (Fig. 3) but it can also develop during the course of disease or even as a manifestation of transformation of an indolent subtype of lymphoma to a higher grade, most commonly diffuse large B-cell lymphoma (Figs. 5 and 9). Overall, the prognosis of lymphomatous transformation to higher grades is generally poor[6].

Use of FDG-PET/CT

Although CT is considered the most convenient imaging technique for the evaluation of patients with lymphoma,

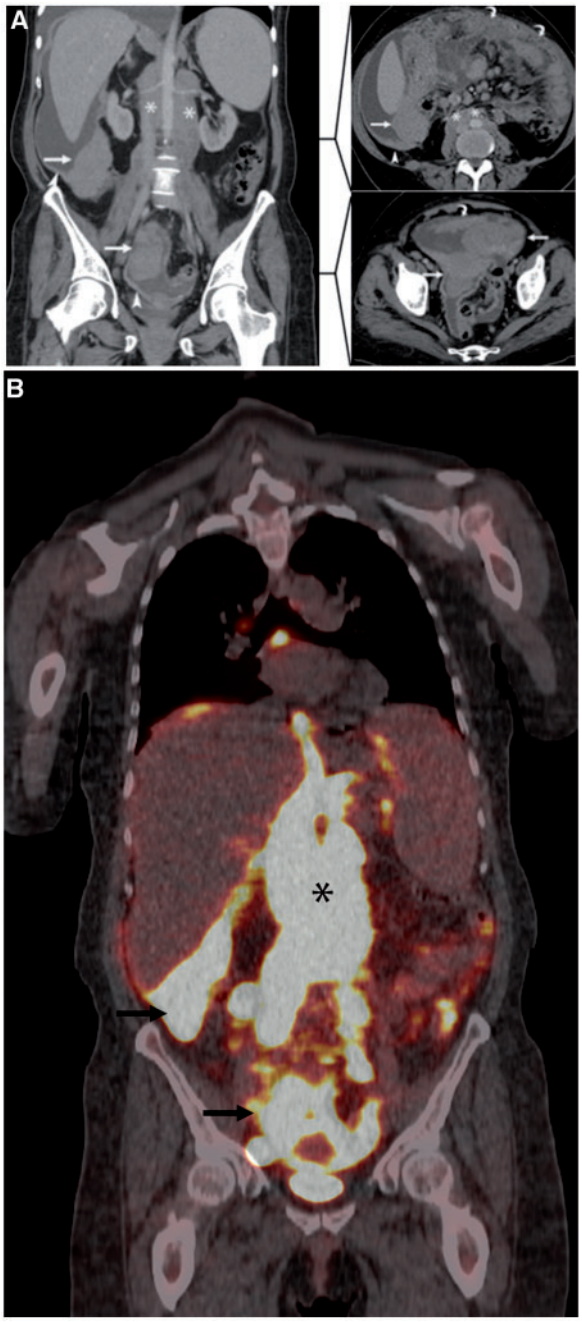

Figure 2 **A 60-year-old woman with recurrent, refractory, diffuse, large B-cell lymphoma who has failed multiple chemotherapy regimens. (A) Reformatted coronal and axial contrast-enhanced CT images show bulky homogeneous masses (arrows) and soft tissue (curved arrows) infiltrating the greater omentum, associated with bulky retroperitoneal lymphadenopathy (asterisks), enlarged mesenteric lymph nodes and moderate ascites. The peritoneum is thickened and shows enhancement (arrowheads). (B) Reformatted coronal fused PET/CT images show intense FDG uptake in retroperitoneal lymphadenopathy (asterisk) and omental masses (arrows).**

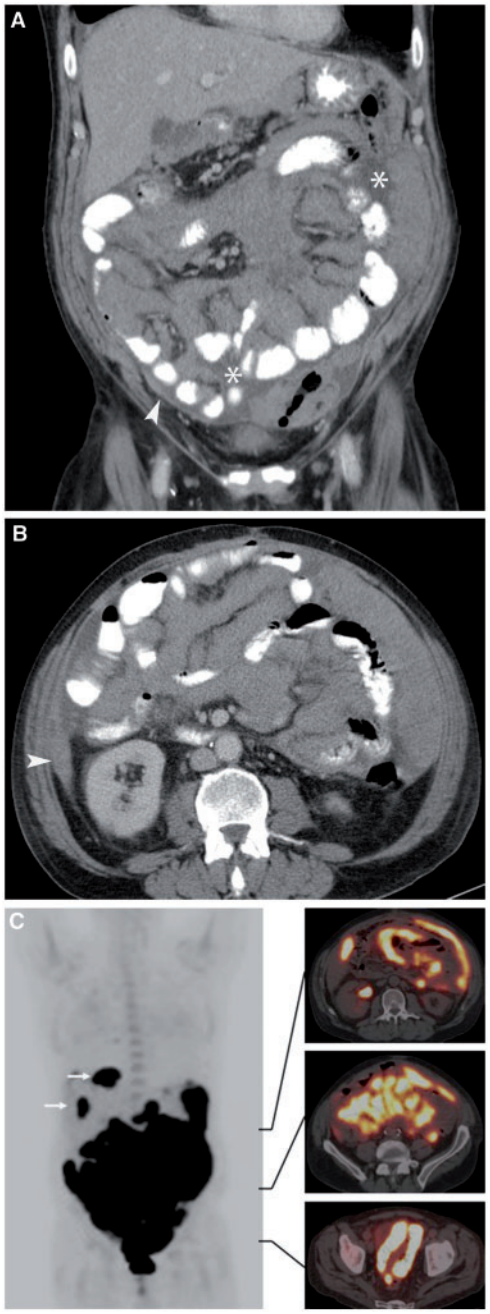

Figure 3 **A 69-year-old-man with diffuse large B-cell lymphoma at the time of diagnosis. Reformatted coronal (A) and axial (B) contrast-enhanced CT images show diffuse homogeneous soft tissue infiltration along leaves of mesentery, associated with peritoneal thickening (arrowheads, A, B) and mild ascites (asterisks, A). Coronal maximum intensity projection PET image and axial fused PET/CT images (C) show diffusely increased metabolic activity in the omentum, mesentery and peritoneal lining, consistent with extensive lymphomatous involvement. Bowel involvement was suspected on conventional contrast-enhanced CT, however axial fused PET/CT images show no FDG uptake in the small or large bowel. Note the FDG-avid cardiophrenic and epiphrenic enlarged lymph nodes (arrows, C).**

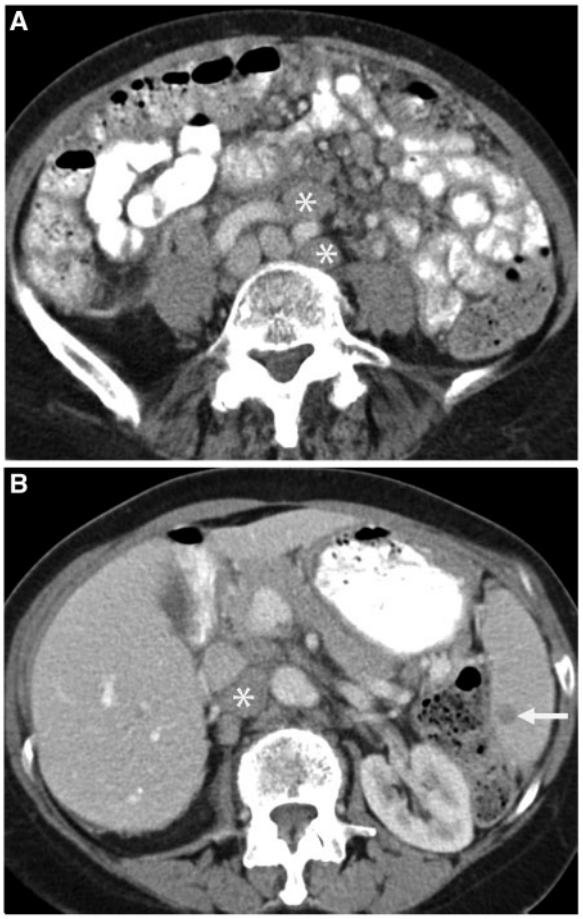

Figure 4 **A 67-year-old man with diffuse large B-cell lymphoma. (A) Axial contrast-enhanced CT image shows multiple small peritoneal nodules and a smudged appearance caused by the infiltrated adjacent omental fat. Retroperitoneal lymphadenopathy is also seen (asterisks). (B) Axial contrast-enhanced CT image shows additional retroperitoneal enlarged lymph nodes (asterisk) and lymphomatous involvement of the spleen (arrow).**

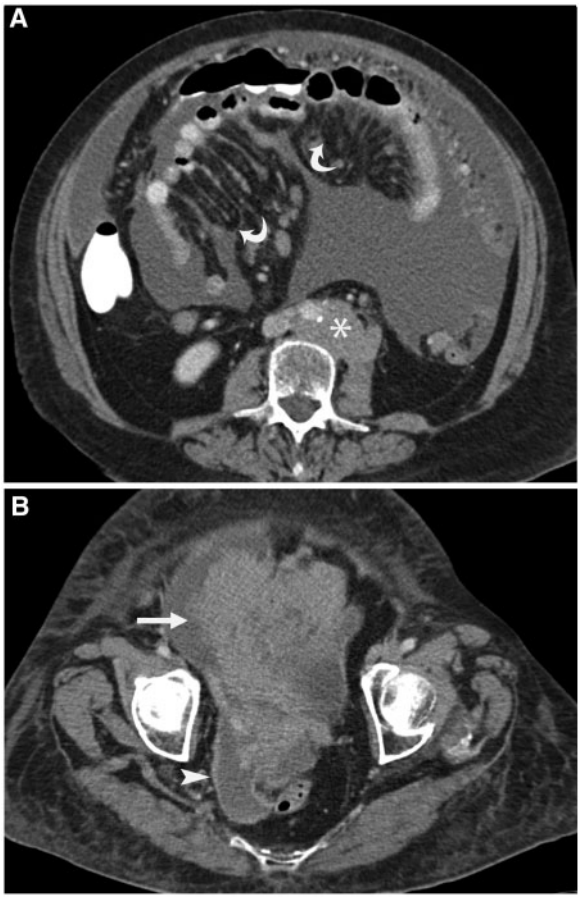

Figure 5 **A 71-year-old woman with a history of follicular lymphoma with transformation to a diffuse large B-cell lymphoma. (A) Axial contrast-enhanced CT images shows prominent straightened vessels secondary to lymphomatous infiltration of the mesentery causing a stellate mesenteric appearance (curved arrows) associated with large volume ascites. Retroperitoneal lymphadenopathy is also seen (asterisk). (B) Axial contrast-enhanced CT image shows bulky homogeneous peritoneal masses (arrow) within the pelvis associated with free fluid and peritoneal enhancement (arrowhead).**

FDG-PET/CT has been shown to improve staging, evaluation of therapy response and earlier detection of recurrent disease. Due to its improved usefulness in distinguishing residual active lymphoma and benign fibrosis, FDG-PET/CT has been found valuable in assessment of treatment response (Fig. 8). FDG-PET/CT is also helpful in the selection of suitable sites for biopsy, because this imaging modality not only demonstrates sites that are accessible but also that the site of interest is metabolically active, increasing the likelihood of a diagnostic result (Fig. 7). Assessment of prognosis is very important for therapy decisions. The International Prognostic Index used for NHL is based on both imaging and clinical findings, and the stage and the number of extranodal sites of disease are included. In Hodgkin lymphoma, these parameters are also of critical importance for the management strategies[6]. In peritoneal lymphomatosis, FDG-PET/CT not only has greater sensitivity than CT for depicting peritoneal involvement in subtle cases but it also has greater accuracy in identifying extranodal sites other than the peritoneum, even in the absence of CT findings (Fig. 7). The FDG-PET/CT findings modify the treatment approach in approximately 25% of patients[8]. Published data have been encouraging and suggest that FDG-PET/CT has a higher sensitivity in comparison with multidetector CT to detect peritoneal malignancies in general. The findings from retrospective studies have demonstrated that FDG-PET/CT has a sensitivity as high as 100% and a specificity as high as 97% for the detection of peritoneal seeding, compared with a sensitivity of 88% and a specificity of 97% for multidetector CT[9].

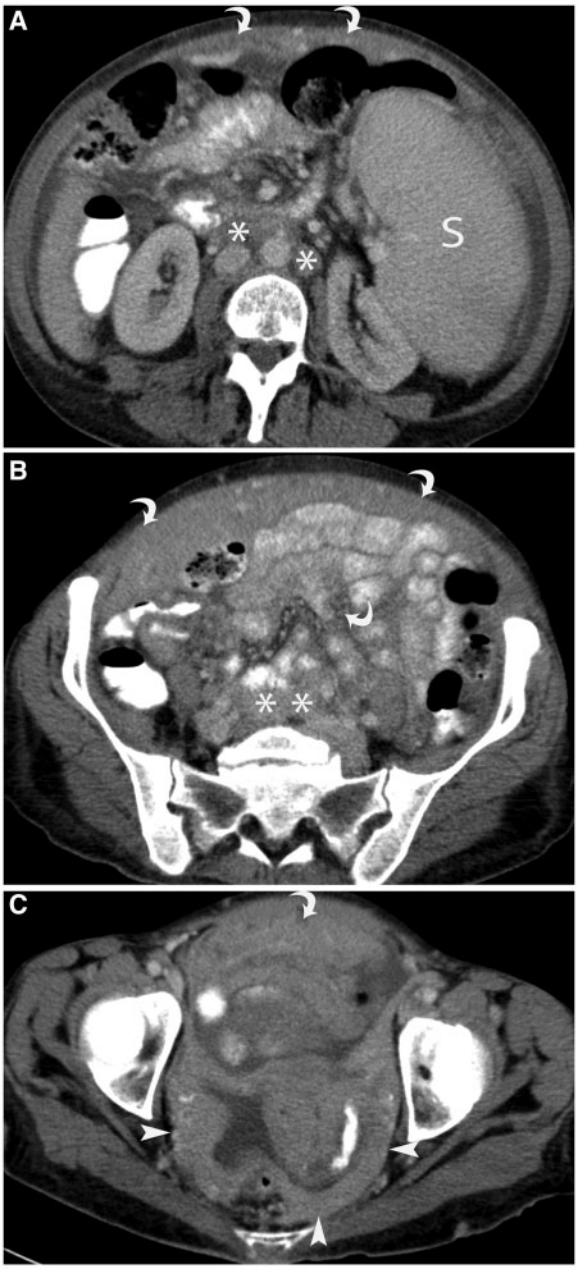

Figure 6 **A 54-year-old woman with a refractory mantle cell lymphoma. Axial contrast-enhanced CT images (A–C) show homogeneous soft tissue in the greater omentum (curved arrows, A–C) and along the leaves of the mesentery (curved arrow, B) associated with marked peritoneal thickening (arrowheads, C) and mild ascites. Moderate splenomegaly (letter S, A) and multiple enlarged retroperitoneal lymph nodes (asterisks, A, B) are also seen.**

There are 2 distinct patterns of glucose metabolism on FDG-PET/CT in peritoneal lymphomatosis: a nodular (Figs. 7 and 8) and a diffuse (Figs. 2 and 3) metabolic pattern. The FDG avidity in lymphoma is variable. Aggressive and highly aggressive NHL and Hodgkin lymphoma usually show high FDG avidity. However, indolent lymphomas have overall low-grade or no FDG uptake because of its low metabolic activity. In patients with FDG-avid lymphomas, a complete metabolic response after therapy is a good indicator that the patient will remain disease-free in the long term. FDG avidity after 2–4 cycles of chemotherapy has been related to poor clinical outcome. In indolent lymphomas, treatment response is usually measured taking symptom relief, overall survival and progression- and event-free survival into consideration. FDG-PET/CT is valuable in cases where transformation of an indolent lymphoma to a higher grade subtype is suspected, by depicting abnormal FDG avidity at sites of transformation, because indolent lymphomas usually have low-grade FDG uptake (Fig. 9). Some subtypes of lymphoma (e.g., extranodal marginal zone lymphoma) show variable FDG uptake and a baseline scan for comparison is of utmost importance in the assessment of therapeutic response. In most cases, FDG-PET/CT should be performed 6–8 weeks after conclusion of therapy to minimize misleading FDG uptake due to post-treatment inflammation[6].

Distinguishing peritoneal lymphomatosis from peritoneal carcinomatosis and peritoneal sarcomatosis

The diagnosis of peritoneal lymphomatosis can be very challenging because it may simulate peritoneal carcinomatosis or peritoneal sarcomatosis. Peritoneal and omental seeding are known sites of dissemination of metastatic carcinoma, most commonly arising from the ovary, colon and stomach[2,3]. Nevertheless, CT findings of omental caking with homogeneous bulky masses rather than a nodular pattern (Fig. 10), in addition to a diffuse distribution of enlarged lymph nodes, are helpful signs of peritoneal lymphomatosis[4]. In peritoneal carcinomatosis, lymphadenopathy is usually located around the primary tumor[2]. Ascites is a frequent finding in carcinomatosis, and may be moderate to large volume. Although bulky masses are also usually seen in peritoneal sarcomatosis, they are frequently heterogeneous, hypervascular and may be associated with hemoperitoneum (Fig. 11)[10]. Furthermore, lymph node involvement in sarcoma is very rare. A prospective evaluation of a large group of patients with soft tissue sarcoma found a prevalence of 2.7% for lymph node metastasis[11].

Conclusion

The appearance of peritoneal lymphomatosis may overlap with carcinomatosis and sarcomatosis, however bulky homogeneous masses or smooth peritoneal soft tissue thickening, diffuse lymphadenopathy, in addition to imaging features of variable extranodal lymphomatous involvement are contributory findings supporting the lymphoma diagnosis. FDG-PET/CT is a valuable asset

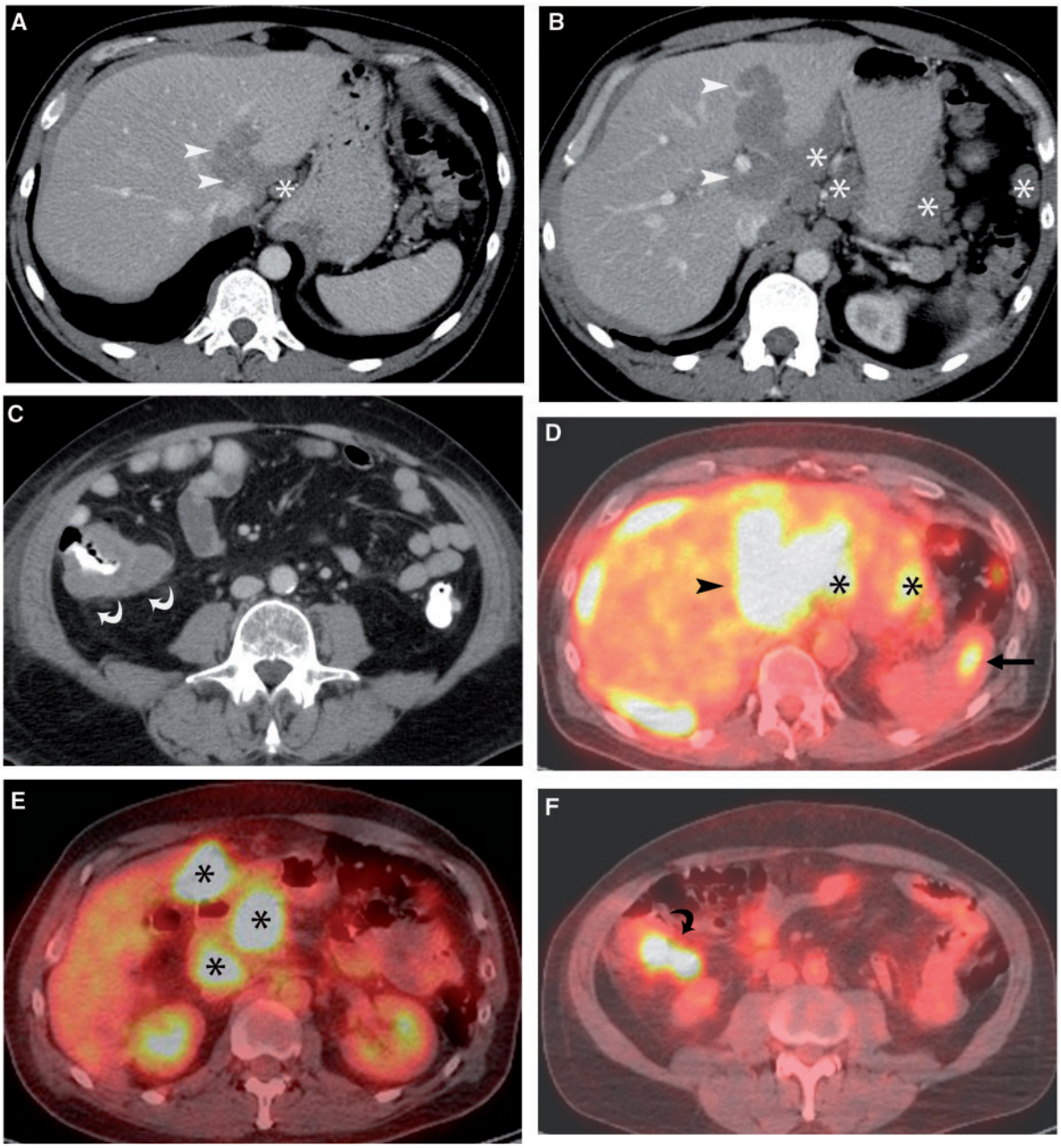

Figure 7 **A 50-year-old man with refractory follicular lymphoma. Axial contrast-enhanced CT images (A–C) show lymphomatous infiltration in the periportal region (arrowheads, A, B), with nodules and masses in the lesser sac and greater omentum (asterisks, A, B). Cecal wall thickening (curved arrows, C) is also noted, representing lymphomatous involvement. Axial fused PET/CT images (D–F) show intense FDG uptake by the soft tissue infiltrating the periportal region (arrowhead, D), the peritoneal nodules (asterisks, D, E), and the cecum (curved arrow, F). A focal area of FDG uptake in the spleen is consistent with lymphoma involvement (arrow, D) and was not seen in the contrast-enhanced CT (A). Despite the intense FDG uptake, a biopsy did not show evidence of transformation to a diffuse large B-cell lymphoma.**

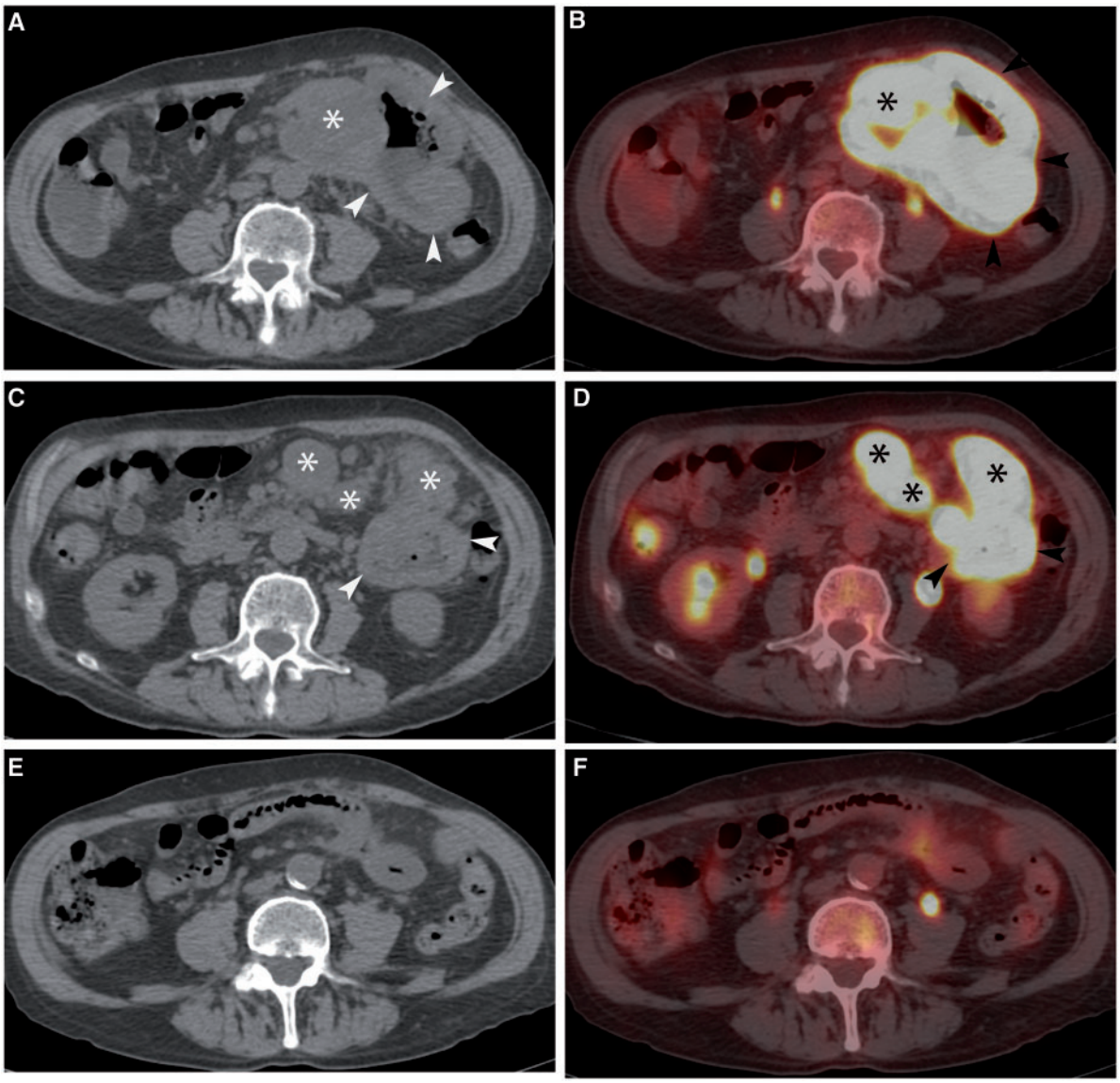

Figure 8 **A 65-year-old man with Burkitt lymphoma and complete response to treatment after 6 cycles of chemotherapy. Axial CT images (A, C) and axial fused PET/CT (B, D) images show aneurysmal dilatation and thickening of a jejunal segment with intense FDG uptake (arrowheads, A–D) associated with nodular FDG uptake in the omentum that corresponds to large omental nodules and masses (asterisks, A–D). Axial CT image (E) and axial fused PET/CT image (F) show complete resolution of the jejunal thickening (E) and no evidence of residual FDG uptake (F) after treatment.**

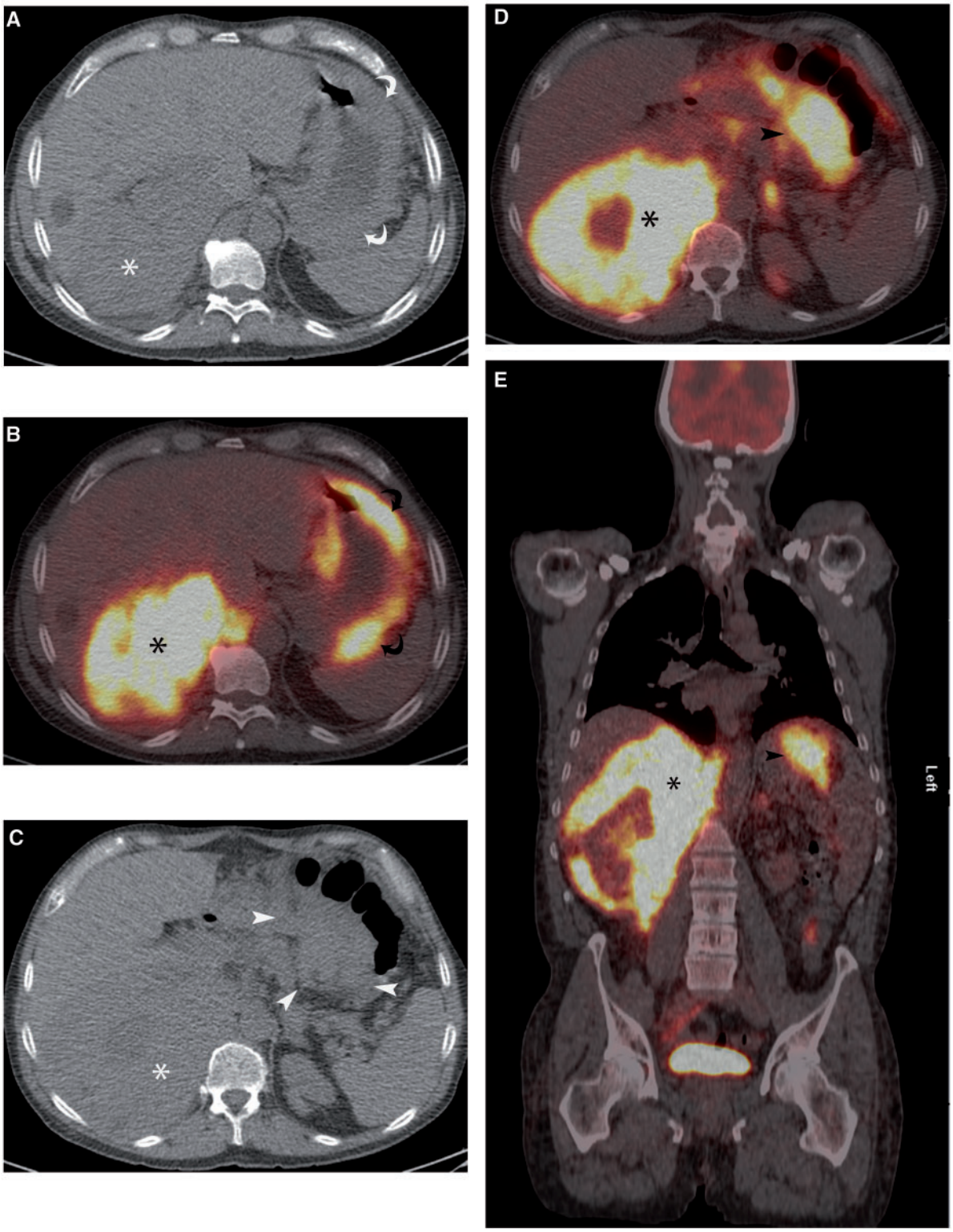

Figure 9 **A 58-year-old man with an 8-year history of follicular lymphoma with transformation to a diffuse large B-cell lymphoma. Axial CT images (A, C) and axial (B, D) and coronal (E) fused PET/CT images show mild diffuse thickening of the gastric wall with intense FDG avidity (curved arrows, A, B) associated with an adjacent homogeneous soft tissue mass in the greater omentum showing intense FDG uptake consistent with peritoneal lymphomatosis (arrowheads, C–E). There is also right perirenal tissue with intense FDG uptake consistent with lymphomatous involvement (asterisks, A–E). Transformation was suspected secondary to intense FDG uptake, which was ultimately proven by biopsy of the perirenal tissue.**

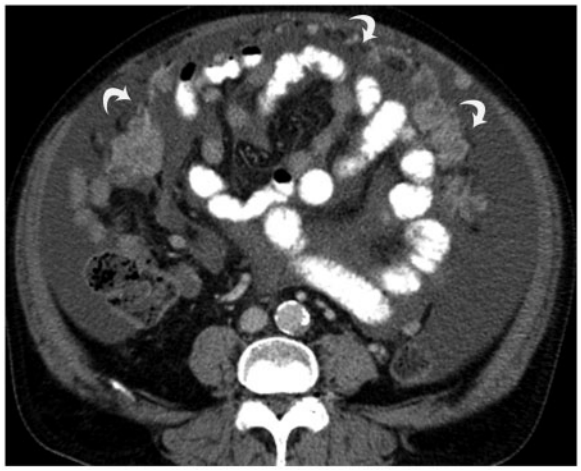

Figure 10 **A 49-year-old woman with ovarian cancer and peritoneal carcinomatosis. Axial contrast-enhanced CT image shows small soft tissue nodules (curved arrows) within the greater omentum associated with large volume ascites. There is no retroperitoneal lymphadenopathy.**

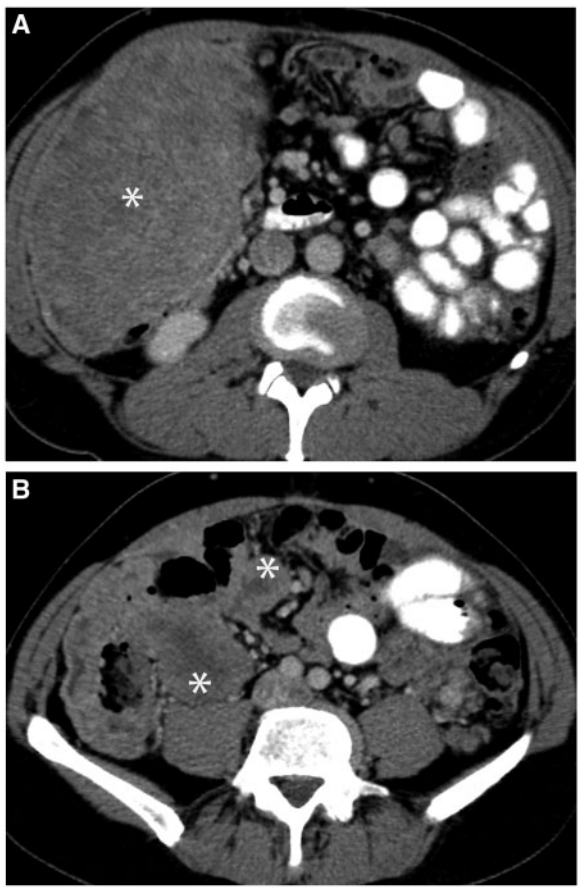

Figure 11 **A 52-year-old man with a gastrointestinal stromal tumor and peritoneal sarcomatosis. Axial contrast-enhanced CT images (A, B) show bulky heterogeneous masses within the greater omentum (asterisks, A, B) with no associated ascites or enlarged lymph nodes.**

in the staging and evaluation of metabolic response to therapy, therefore optimal management of the disease requires that the radiologist be familiar with the role of FDG-PET/CT, because its findings may influence treatment decisions.

Conflict of interest

The authors have no conflicts of interest to declare.

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