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CT in Pseudomyxoma Peritonei: A Review of 17 Cases

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AIM: To describe the computed tomography (CT) findings in pseudomyxoma peritonei.
MATERIALS AND METHOD: Two observers independently and retrospectively reviewed the CT images of 17 consecutive patients (nine women, eight men, mean age 53 years) with histologically proven pseudomyxoma peritonei.
RESULTS: Six patients had small volume disease where pseudomyxoma peritonei was present in focal collections in the peritoneal cavity. Eleven had large volume disease that completely, or almost completely, filled the peritoneal cavity. Pseudomyxoma peritonei is characterized by low attenuation mucinous ascites on CT. Areas of high attenuation, septae and calcification are seen more commonly within it as the volume of disease increases. The pattern of accumulation of pseudomyxoma peritonei follows the normal flow of peritoneal fluid. It initially seeds at sites of relative stasis and as large volume disease develops it fills the remaining spaces in the peritoneal cavity and pressure effects dominate imaging. Pseudomyxoma peritonei may extend into hernial orifices or the pleural cavity.
CONCLUSION: Pseudomyxoma peritonei is difficult to diagnose clinically. However, the pattern of accumulation of disease is predictable and can be recognized on CT. Sulkin, T. V. C. *et al.* (2002). *Clinical Radiology* **57**, 608–613.

Key words: pseudomyxoma peritonei, computed tomography, mucinous ascites.

Pseudomyxoma peritonei (PP) is characterized by the gradual accumulation of large volumes of mucinous ascites in the peritoneal cavity. There is now a convincing body of evidence that it arises from a ruptured mucin-producing tumour of the appendix [1–3]. PP is distinct from disseminated mucinous carcinomatosis, a condition characterized by the accumulation of mucinous ascites secondary to a high-grade mucin producing carcinoma. At either extreme, distinction between the two entities is straightforward, but there is continuing debate as to the point of separation between them [3]. The issue is an important one, as treatment and prognosis differ [1,4].

Historically, surgical treatment of PP has centred on palliative debulking. The aim of modern treatment is to address the relentless re-accumulation of mucinous material. This involves radical surgery to remove all gross disease, supplemented with perioperative intraperitoneal chemotherapy [1,3]. In view of the shift in the treatment goal from palliation to possible cure, it is important to make the diagnosis of PP as early as possible so as to expedite referral to a specialist centre. Clinical presentation is non-specific. Suspected acute appendicitis, increasing abdominal girth or a new onset hernia are the commonest presenting symptoms [5]. Many patients will undergo computed tomography (CT) as part of their diagnostic work-up. Given that the pattern of spread of disease in PP is predictable, and this can be recognized on CT, the radiologist is in a unique position to suggest the diagnosis.

We present a review of the CT features of PP based on the CT findings of 17 patients referred for surgical cytoreduction.

METHOD

The hard-copy abdominal and pelvic CT images of 17 consecutive patients (nine women and eight men, mean age 53 years) referred for surgical cytoreduction with histologically proven PP were independently and retrospectively reviewed by two observers. Where there was disagreement a consensus opinion was reached. Patients without a histological diagnosis were excluded. The CT examinations were performed between October 1995 and February 2000.

The images were assessed for the presence or absence of PP in the following areas: the right and left subphrenic spaces and paracolic gutters, Morison's pouch, the pouch of Douglas/rectovesical pouch and the porta hepatis. The costophrenic recesses on the images through the upper abdomen and the hernial orifices were reviewed. PP adjacent to the liver, spleen, stomach, duodenum, jejunum or ileum, the transverse and sigmoid colon and around the small bowel mesentery was also recorded.

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Table 1 – The CT distribution of pseudomyxoma peritonei and visceral involvement in small volume disease (SVD)

Distribution of PP/visceral involvement	Percentage of cases ($n = 6$)
Right subphrenic space	67% (4)
Surface of liver	67% (4)
Left subphrenic space	67% (4)
Surface of spleen	67% (4)
Morison's pouch	33% (2)
Right paracolic gutter	50% (3)
Left paracolic gutter	17% (1)
Pouch of Douglas/rectovesical pouch	100% (6)
Jejunum or ileum	17% (1)
Bowel obstruction	0
Hernial orifices	0
Thorax	0

Table 2 – The CT features of pseudomyxoma peritonei in small volume disease (SVD)

CT features	Percentage of cases ($n = 6$)
Visceral scalloping	100% (6)
Septae in PP	67% (4)
Calcification in PP	0
Right ovarian mass	40% of women (2)
Appendiceal tumour	17% (1)

The presence or absence of the following features were noted: visceral scalloping, septae or calcification in the mucinous material, displacement of the small bowel posteriorly or centrally, bowel obstruction, an appendiceal tumour and an ovarian mass.

The CT images were acquired on a GE Pace CT system (General Electric, Milwaukee, WI, U.S.A.). Oral (1000 ml), rectal (200 ml) and intravenous (150 ml) contrast medium was administered. A slice thickness of 10 mm with an interslice gap of 10 mm or 15 mm was used.

Table 3 – The CT distribution of pseudomyxoma peritonei and visceral involvement in large volume disease (LVD)

Distribution of PP/visceral involvement	Percentage of cases ($n = 11$)
Right subphrenic space	100% (11)
Surface of liver	100% (11)
Left subphrenic space	100% (11)
Surface of spleen	100% (11)
Morison's pouch	82% (9)
Lesser sac	82% (9)
Left paracolic gutter	100% (11)
Right paracolic gutter	100% (11)
Pouch of Douglas/rectovesical pouch	100% (11)
Porta hepatis	82% (9)
Stomach	91% (10)
Duodenum	91% (10)
Jejunum or ileum	100% (11)
Transverse colon	55% (6)
Sigmoid colon	82% (9)
Bowel obstruction	0
Small bowel mesentery	91% (10)
Hernial orifices	27% (3)*
Thorax	9% (1)

*PP found in both inguinal canals extending into the scrotum (1), a paraumbilical hernia (1) and a hiatus hernia (1).

RESULTS

While all the cases in our series had histologically proven PP, they were a mixed group referred with variable volume of disease. Some had undergone a laparotomy at the base hospital before referral. In all patients there was characteristic low attenuation mucinous ascites in the peritoneal cavity. Where large focal collections were present, and in diffuse disease, the appearance was more complex with additional areas of high density within the PP. In order to clarify the CT findings in PP we have divided our cases into two groups:

(1) The first group comprises those with small volume disease (SVD) present in focal collections in the peritoneal cavity (six patients, one man, five women, mean age 49 years). The findings are summarized in Tables 1 and 2. All patients with SVD went on to have a complete surgical cytoreduction of PP.
(2) The second group comprises those patients with large volume disease (LVD) completely or almost completely, filling the peritoneal cavity (11 patients, seven men, four women, mean age 54 years). The findings are summarized in Tables 3 and 4. Three patients in the LVD group went on to have a complete cytoreduction of PP, four had an incomplete cytoreduction (two due to small bowel involvement and two due to extensive disease in the upper abdomen) and four were inoperable (three due to small bowel involvement and one due to extensive disease in the upper abdomen).

DISCUSSION

The description of the findings on CT in PP in the radiological literature is limited to case reports and small series [6–16]. The largest imaging series of 33 patients with PP does not specify how many had CT [12]. Although our cases are a heterogeneous group, they have histologically proven PP and illustrate the CT features well.

On CT, PP appears of low attenuation, but when large volumes accumulate, areas of high attenuation can be seen

Table 4 – The CT features of pseudomyxoma peritonei in large volume disease (LVD)

CT features	Percentage of cases ($n = 11$)
Small bowel displaced posteriorly	27% (3)
Small bowel displaced centrally	45% (5)
Visceral scalloping	100% (11)
Septae in PP	100% (11)
Calcification in PP	36% (4)
Left ovarian mass	0
Right ovarian mass	0
Appendiceal tumour	0

within it. This may be due to solid elements within the mucinous material, or compressed mesentery (Fig. 1). We cannot comment on enhancement as all the images in this series were acquired following administration of intravenous contrast medium. Scalloping of visceral surfaces (particularly the liver) is the diagnostic sign that distinguishes mucinous from fluid ascites on CT (Fig. 2) and was identified in all of our cases. Septae (Fig. 3) were identified within the mucinous ascites in all of the LVD cases and in 67% of the SVD group. Non-specific calcification (Fig. 4) was identified in 36% of the LVD patients, but was not seen in SVD. There are therefore no characteristic CT features that distinguish the individual deposits of PP from other causes of mucinous ascites. The pattern of accumulation of disease is, however, characteristic and should suggest the diagnosis.

The circulation of peritoneal fluid in the peritoneal cavity is dictated by gravity, pressure changes associated with respiration and the physical boundaries of the peritoneal reflections [17]. The mucin producing cells in PP are poorly adherent and circulate with peritoneal fluid. They are easily dislodged from the surface of the bowel by constant peristalsis. They only seed at sites of relative stasis. This process has been termed the redistribution phenomenon [1].

The SVD group in our series represents the early stages of the disease before the peritoneal cavity has been filled with PP (Fig. 5). The pouch of Douglas/rectovesical pouch (100%), the right and left subphrenic spaces (67%) and the

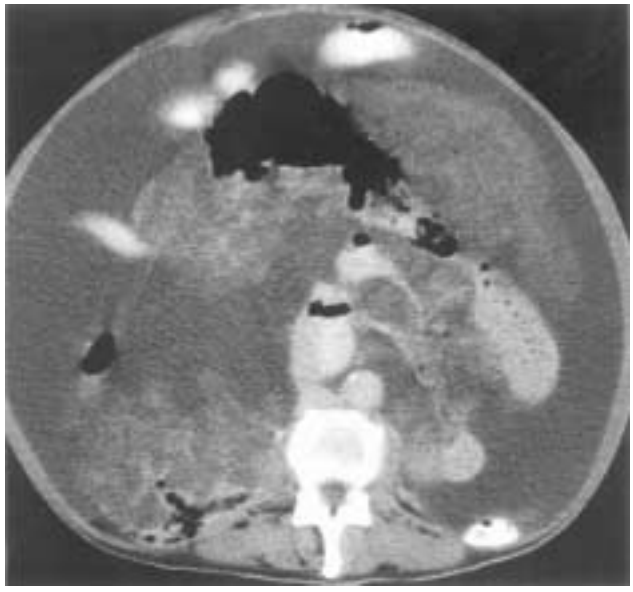

Fig. 1 – High attenuation elements within large volume pseudomyxoma peritonei.

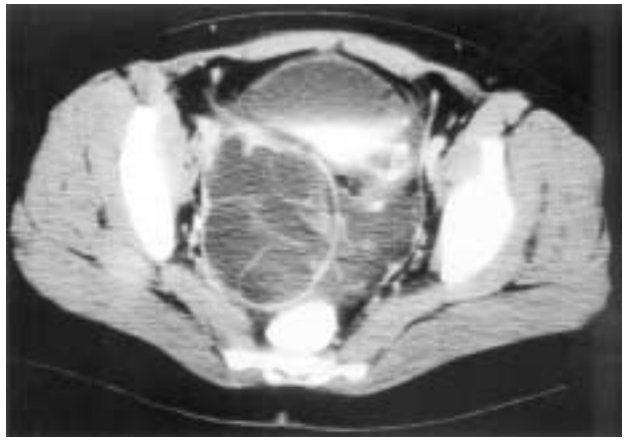

Fig. 3 – Septae within pseudomyxoma peritonei in the pelvis.

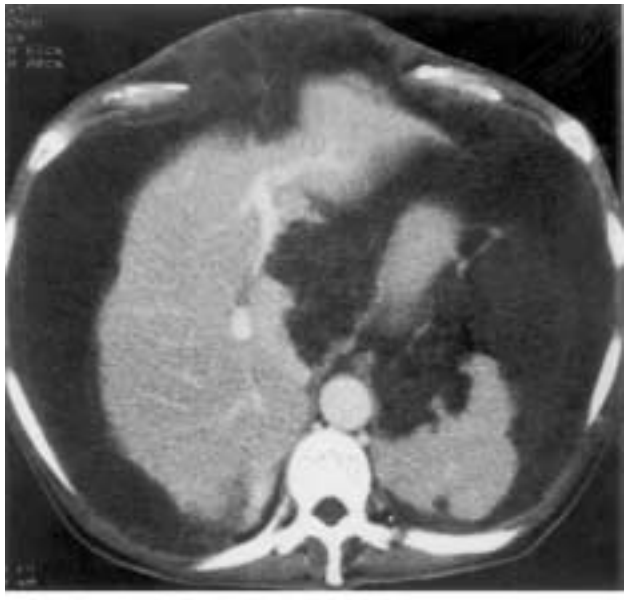

Fig. 2 – Visceral scalloping of the liver and spleen in pseudomyxoma peritonei.

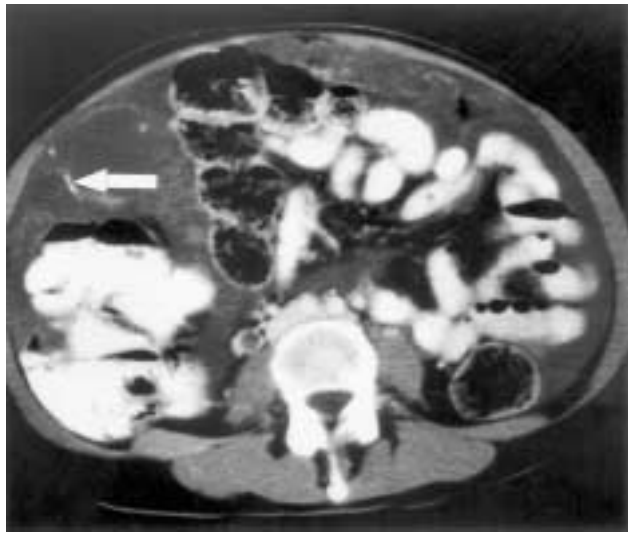

Fig. 4 – Calcification (arrow) in large volume pseudomyxoma peritonei.

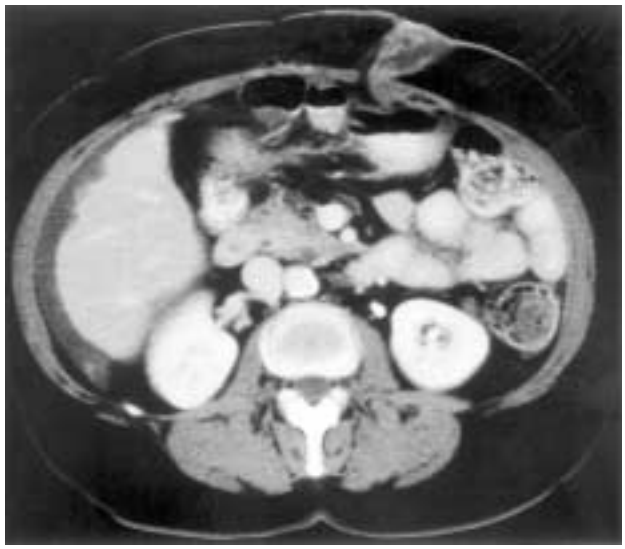

Fig. 5 – Small volume disease in the right subphrenic space causing scalloping of the liver.

surface of the liver and spleen (67%) were the commonest sites involved. This is predictable, as mucin-secreting cells from the ruptured appendiceal tumour will be carried into the pelvis and preferentially up the right paracolic gutter to the right subphrenic space. As the falciform and phrenicocolic ligaments are barriers to involvement of the left subphrenic space, it is surprising that deposits were equally common in both subphrenic spaces and on the surface of the liver and spleen. It is, however, noteworthy that PP was not seen in the left subphrenic space without involvement of the right subphrenic space, and the volume of disease was always greater on the right than the left. Disease in the paracolic gutters was always associated with disease in the respective subphrenic space.

An appendiceal tumour was identified in only one of the SVD group (Fig. 6) and in none of the LVD group (although one had had a previous appendicectomy for an appendiceal mucocele). In large volume disease, the appendiceal tumour is rarely identified among the large volume of mucinous ascites on CT; indeed, it is often difficult to find at operation, as it may be small and fibrosed [1]. In the SVD group, it is surprising that an appendiceal tumour was not more commonly identified. In a recent article Zissin *et al*. described the CT features in 10 cases of mucocele of the appendix; PP was found in 50% [13].

Synchronous mucinous ovarian tumours are reported as occurring in up to 44% of cases of PP in women. They are bilateral in up to 80% [2]. There has been much debate as to whether the primary tumour lies in the appendix or the ovary. The general consensus, backed up by immunohistochemical studies, is that a mucin-producing tumour of the appendix ruptures and this then seeds the ovaries by transcoelomic spread [1–3,18]. A unilateral ovarian mass was identified in two of the five women in the SVD group (40%), but none of the four women in the LVD group. It should be noted that one of the LVD patients had undergone previous surgery for a left ovarian cystadenoma and appendiceal mucocele.

Once PP fills the sites listed above, it involves those sites where peristalsis is limited by peritoneal attachments and finally goes on to fill the remainder of the peritoneal cavity. Although the mucin-producing cells remain poorly adherent, the large volume of disease can overcome peristalsis and indent the small bowel. This is only a feature of advanced disease [1]. As demonstrated in this series, small bowel obstruction is not a feature of PP. This is due to the fact that the tumour cells do not invade the visceral peritoneum [1].

In the LVD cases the peritoneal cavity was completely, or almost completely, filled with PP (Fig. 1). At this stage the characteristic pattern of spread of PP cannot be appreciated and it is hard to distinguish PP from other causes of mucinous ascites. PP may fill paraumbilical, inguinal and hiatus herniae (Figs 7 and 8). Large deposits of PP may

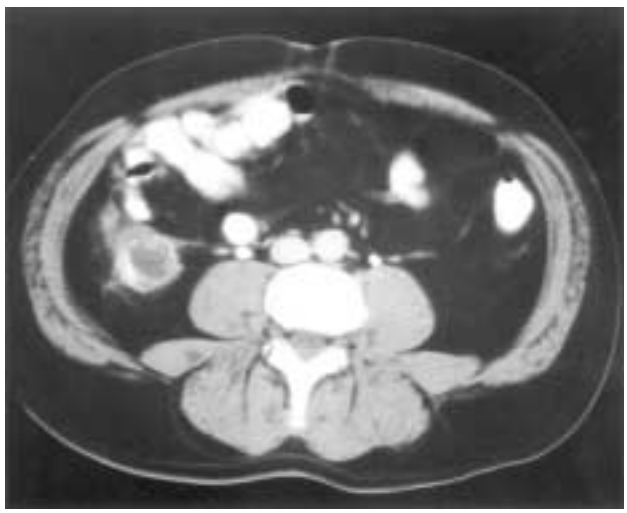

Fig. 6 – Appendiceal tumour in a case of pseudomyxoma peritonei.

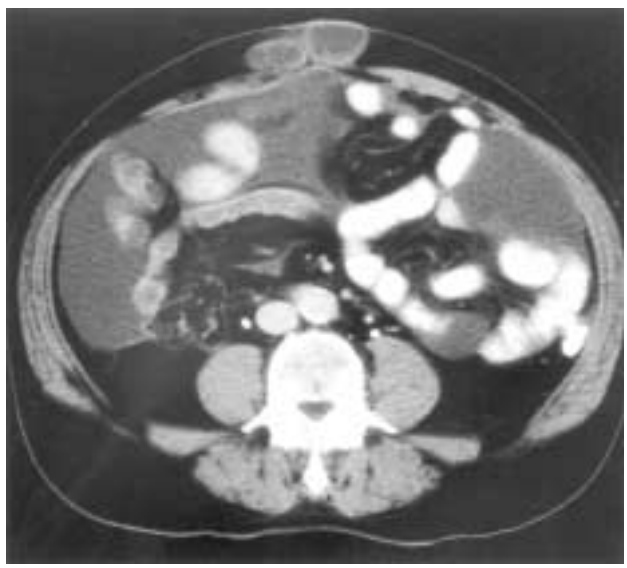

Fig. 7 – Pseudomyxoma peritonei in a paraumbilical hernia.

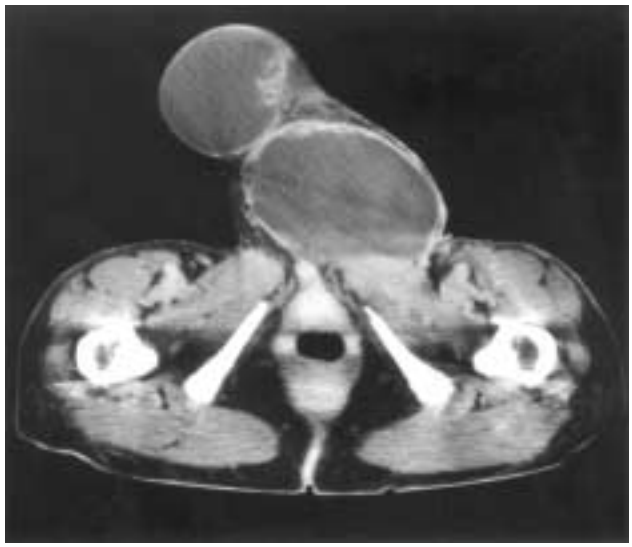

Fig. 8 – Pseudomyxoma peritonei in both sides of the scrotum in a patient with bilateral inguinal herniae.

invaginate into the liver and spleen thus mimicking parenchymal deposits (Fig. 9). No hepatic or splenic metastases were identified at operation in our series and they are not a feature of PP described elsewhere [1].

PP was identified surrounding the loops of jejunum and ileum in all of the LVD cases. In our centre, surgery involves a six-stage peritonectomy. The distal ileum may be resected as part of a right hemicolectomy. Scope for resection of the more proximal ileum and the jejunum is limited. Deposits of PP can be removed from the small bowel, but this can be technically difficult, particularly if there has been previous surgery. Complete removal of tumour may therefore be impractical where there is extensive small bowel involvement [1,19]. It is noteworthy that a study that evaluated the ability of pre-operative CT to predict the likelihood of a complete surgical cytoreduction in 45 patients with mucinous peritoneal carcinomatosis (including 10 cases of PP) found that volume of disease does not predict surgical outcome. The only factors that were prognostic were bowel obstruction and tumour deposits on the jejunum and proximal ileum [19]. Given the small numbers in our series, the retrospective nature of the review and the fact some patients had had previous surgical intervention, it is difficult to draw conclusions regarding the CT features that predict a complete surgical cytoreduction. It would appear that SVD is a good prognostic sign (100% complete cytoreduction). Excessive disease in the upper abdomen and small bowel involvement identified at operation were the causes of irresectability or incomplete surgical cytoreduction. Reliable identification of these latter features on CT is difficult. Our study was limited by the use of a conventional CT system and we would advocate using helical or multi-slice CT where possible. Further prospective studies using CT or magnetic resonance imaging (MRI) are required to establish the imaging features that are prognostically important.

Pressure effects were a prominent feature in the LVD group. Both central (45%) and posterior (27%) small bowel displacement were seen (Fig. 10). The abdomen was universally distended and the retroperitoneal structures compressed (Fig. 10). Neither abdominal nor pelvic lymphadenopathy was a feature in any of our cases. PP is not characterized by haematogenous or lymphatic metastases, so the presence of lymphadenopathy should bring the diagnosis of PP into question.

One patient with large volume disease, who had not had previous surgery, demonstrated unilateral pleural thickening. In the clinical context and given the absence of an alternative aetiology, these were assumed to be pleural deposits of PP (Fig. 11). Pleural PP is an unusual finding

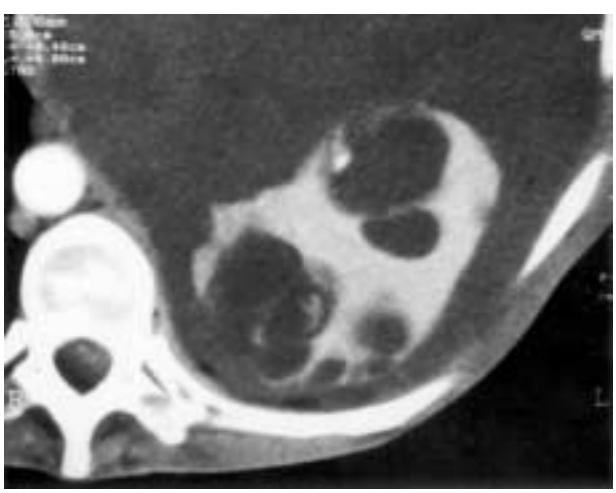

Fig. 9 – Pseudomyxoma peritonei invaginating into the spleen simulating parenchymal deposits.

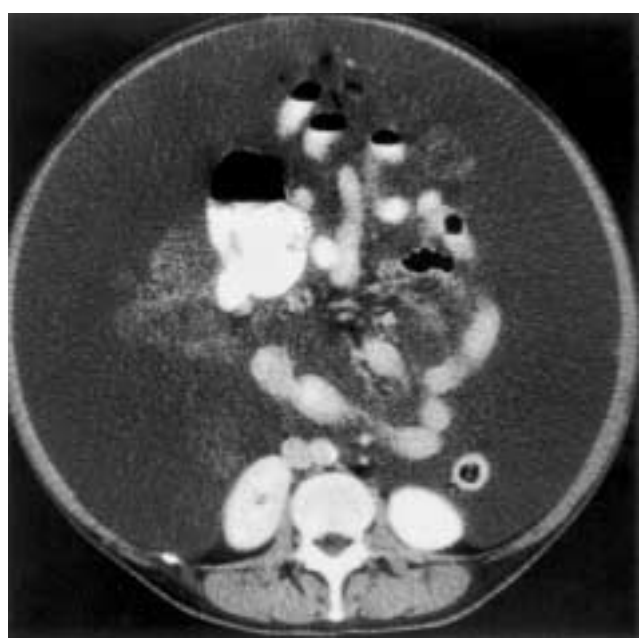

Fig. 10 – Central displacement of the small bowel and compression of retroperitoneal structures by pseudomyxoma peritonei.

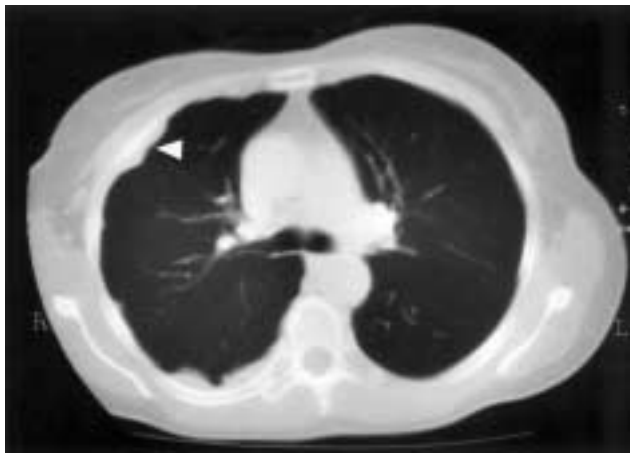

Fig. 11 – Pleural deposits in the right pleural space in a patient with large volume disease (arrowhead). These were assumed to be deposits of pseudomyxoma peritonei (see text).

and is presumably due to the presence of a congenital pleuroperitoneal communication [20], or direct infiltration through the diaphragm [21]. Spread of mucinous material into the pleural cavity is more common in cases of post-operative recurrence [21].

We have reviewed the CT findings in PP. In early disease it is the predictable pattern of accumulation of mucinous ascites, rather than the appearance of the individual deposits, that can point the radiologist towards suggesting a diagnosis of PP. Once disease completely, or almost completely, fills the peritoneal cavity, the CT findings are more non-specific. Further work is required to predict surgical outcome from pre-operative imaging.

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