Case reports

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Pseudomyxoma peritonei

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Pseudomyxoma peritonei is an infrequently reported condition which occurs most commonly in women in the 50–60 year age range although it is well recognized in men. We report a case which illustrates the importance of making a preoperative diagnosis, whilst highlighting the fact that pseudomyxoma is a blanket term for a spectrum of conditions that behave very differently.

Case report

The patient, a 74-year-old Polish man, presented complaining of ankle swelling associated with mild calf ache. It was only on direct questioning that he acknowledged that his abdominal girth had increased considerably over the last five years and that he had put on weight. There were no other symptoms. Until ten years earlier he had drunk half a bottle of spirits a day but now had only the occasional whisky. He smoked half an ounce of tobacco a day.

On examination he was clinically anaemic. The principal finding was of a huge, firm, immobile, irregular mass in the abdomen. Rectal examination revealed only normal stool and a slightly enlarged benign prostate gland. There was no palpable lymphadenopathy. The other positive finding was of mild pitting ankle oedema with overlying reddened skin.

He had a hypochromic anaemia with a haemoglobin of 10.4 g/dl. A blood film showed moderate rouleaux formation with mild aniso- and poikilocytosis. Bone marrow trephine was normal. The erythrocyte sedimentation rate (ESR) was 96 mm in one hour. Apart from a somewhat raised alkaline phosphatase of 137 IU (normal range 30–115 IU), liver function tests were normal. He had slightly increased alpha-1 and alpha-2 globulin levels with normal albumin. Electrolytes and renal function tests were in the normal range.

Plain abdominal X-ray outlined the mass and demonstrated numerous areas of calcification, some of which had an annular pattern. Intravenous urography showed that the mass was pushing the bladder forward over the pelvic brim (Figure 1). Ultrasound scan revealed multiple echogenic areas giving the appearance of multi-septate ascites containing cystic areas. The bowel did not float in the fluid. The liver appeared normal and separate from the abnormal tissue (Figure 2). Computerized tomography confirmed the presence of a dense, fairly homogeneous substance in the abdomen in which the bowels were pushed to one side and which was not typical of ascites. Annular calcification and scalloping of the liver margins were also noted (Figure 3).

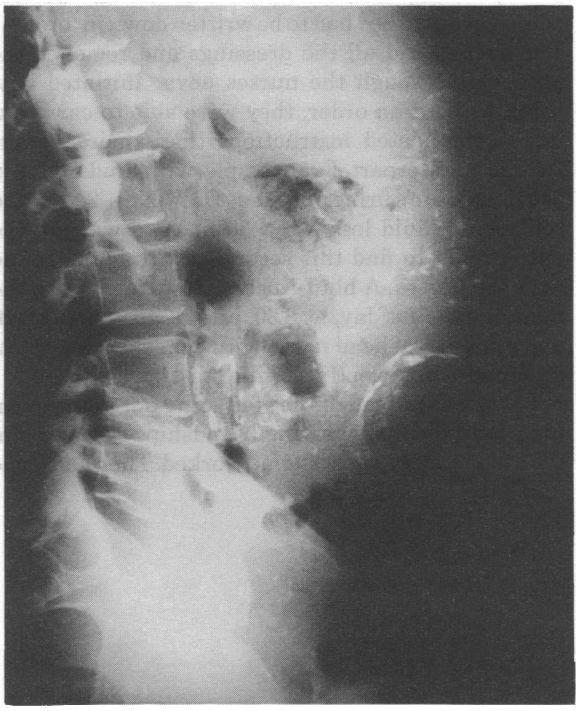

Figure 1. Intravenous urogram showing abdominal mass with annular calcification pushing bladder out of pelvis

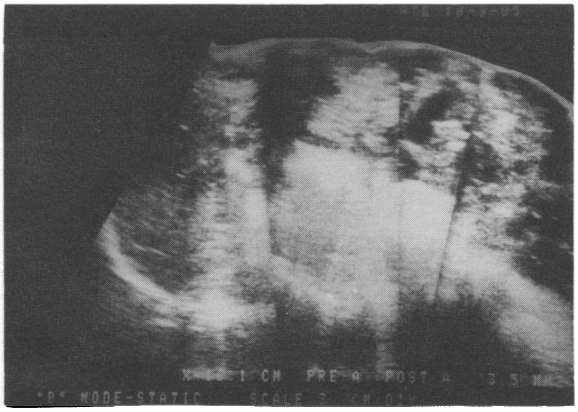

Figure 2. Ultrasound scan showing the appearance of multiseptate ascites but without free-floating bowel

In view of the clinical picture, X-ray, ultrasound and CT findings, a diagnosis of pseudomyxoma peritonei was made. The patient has had no active treatment and is currently being kept under review.

Discussion

Pseudomyxoma peritonei is a condition in which a large quantity of mucinous fluid or a more gelatinous mass accumulates in the abdomen. The nature of the underlying pathological process is conten-

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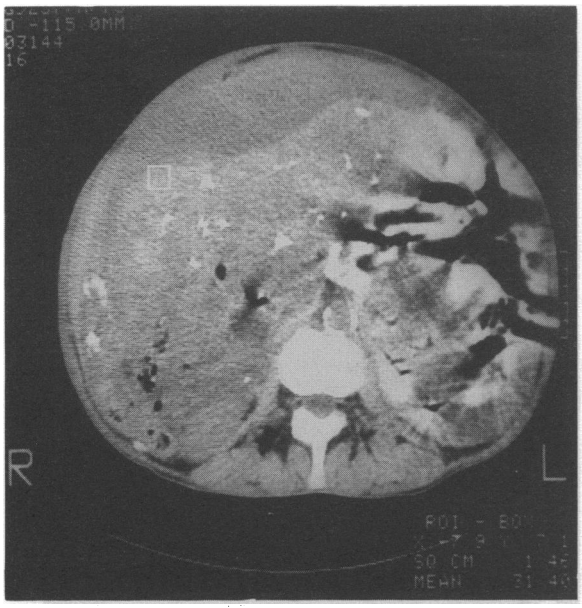

Figure 3. Abdominal CT scan showing the homogeneous mass with annular calcification. Note scalloping of the liver edge

tious. It probably occurs secondary to rupture of a viscus lined with mucin-producing epithelium[1]. The role of secondary implantation of mucin-secreting material and of metaplasia of peritoneal epithelium is unclear. Tumours of the appendix and of the ovary are mainly implicated, but there have been reports associating pseudomyxoma peritonei with mucinous adenocarcinoma of the pancreas[2] and with hyperplastic heterotopic gastric mucosa occurring in a patient with duplication of the ileum[3]. Most ovarian tumours are cystadenocarcinomas but underlying mucinous cystadenomas have been found[4]. Conversely, benign disease of the appendix is more common, although pseudomyxoma may arise from appendiceal adenocarcinomas[5,6]. It is this broad spectrum of disease which makes the comparison of cases of pseudomyxoma so difficult, and each new one requires an individual approach. The use of a single term in this context has been criticized[7].

Presentation is indicative of the underlying cause. Increasing abdominal distension in an otherwise well patient, as occurred in the case reported here, is typically associated with pseudomyxoma originating in the appendix[8]. The patient gave no past history suggesting that he may have had an appendiceal abscess, a possible precursor of pseudomyxoma[5,6]. He also had none of the other recorded symptoms which include vague abdominal pain, fever, weight loss to the point of cachexia and those of intestinal obstruction[6,9,10]. Large masses may sometimes give rise to umbilical or other herniae[6,9-11]. Finally, pseudomyxoma may only be discovered at operation[5,6].

The key to diagnosis, which is so important in planning treatment and estimating prognosis, is radiological investigation. Operation is not necessary at this stage. The presence of anaemia[10] and a raised ESR[4,9,12] are well recognized in pseudomyxoma but are not diagnostic. It is the radiological findings already presented in this case that are absolutely characteristic of pseudomyxoma[2,5,10]. Less common is the presence of multiple echogenic spherical masses on ultrasound screening[11].

Views on the treatment of pseudomyxoma are divergent, which probably represents experience with different underlying tumours. It is generally accepted that there is no role for radiotherapy or systemic chemotherapy. Topical chemotherapy using 5-fluorouracil and thiotepa has been tried in an uncontrolled fashion[6,9] and does not seem to confer any benefits. *In vitro* studies suggest that enzymatic digestion using hyaluronidase is not feasible[9]. Other mucolytics have been used unsuccessfully.

The role of surgery is debated. Little and coworkers[9] take the view that the only effective treatment to prolong survival is the removal of the ovary or appendix, as appropriate, together with as much of the mucinous material as possible[9]. In addition, intraperitoneal lavage with 5% dextrose and water has been said to prevent recurrence[4]. The other argument for surgery is that it allows a more accurate estimate of prognosis. The presence of epithelial cells in the acid mucopolysaccharide matrix has been taken as a sign of malignancy[13], while another authority indicates that well differentiated epithelial cells suggest a benign origin[1]. It is to be noted that the distribution of cells may be uneven and that they may be difficult to locate in the pathological specimen.

We feel that surgery is not necessarily indicated. First, some patients have undergone repeated and extensive operations with early demise[6]. Secondly, some patients with pseudomyxoma have survived for more than 15 years[6,8]. It seems likely that the natural history of this type of disease is benign, possibly developing from a benign appendiceal origin. We believe that our patient, whose mass has been slowly enlarging over the last five years, has such a form of pseudomyxoma peritonei. Most importantly he feels well and is not troubled by his abdomen and is dogmatic that he will not have an operation. He can be followed in the outpatient department and should the mass begin to impose mechanical limitations on him we will be able to offer help. This is the only definite indication for surgery within this clinical picture and confirms the importance of making the diagnosis by radiological investigation.

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Aggressive polyfibromatosis

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We report an unusually aggressive case of polyfibromatosis associated with an erosive arthropathy. This association has not previously been described.

Case report

A 48-year-old West Indian man was well until February 1983 when he noticed stiffness of both hands and painful curvature of his penis on erection. In August 1983 he developed bilateral fibrotic palmar contractures. At the same time he noticed several spontaneous skin swellings on his arms and chest.

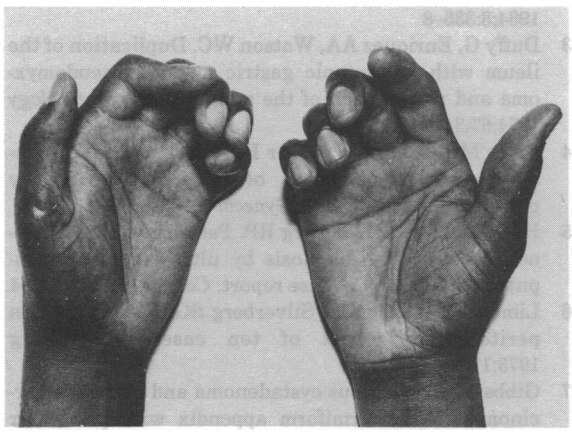

Figure 1. Bilateral palmar fibromatosis with flexion deformities

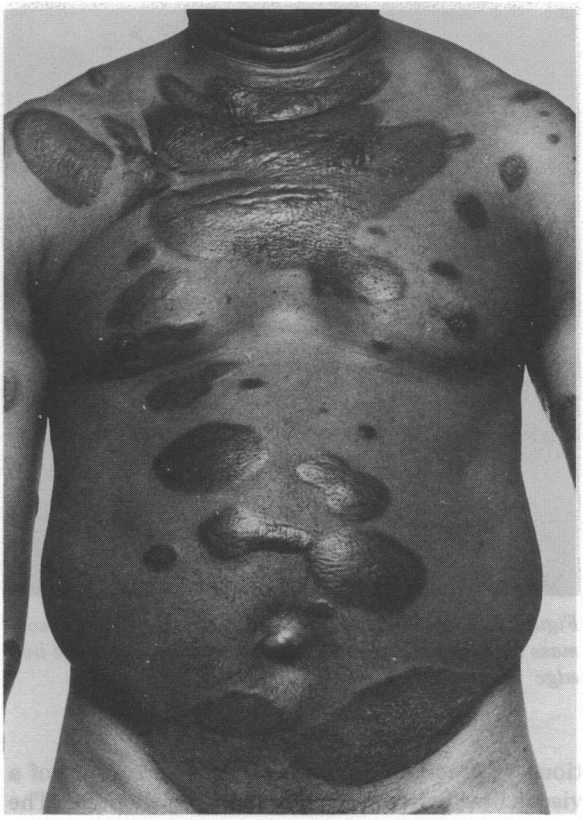

Figure 2. Keloid nodules and plaques

On examination, he had multiple keloidal nodules on his shoulders, arms and presternal area, diffuse fibrotic palmar fibromatosis bilaterally (Dupuytren's contracture) and a fibrotic nodule palpable in the shaft of his penis (Peyronie's disease).

Since then his hands have deteriorated rapidly, producing bilateral flexion deformities (Figure 1) and limited finger movements, so that he is now unable to work or even dress himself. The keloid-like nodules and plaques have continued to enlarge dramatically (Figure 2) and arise both spontaneously and at sites of trauma such as venesection, although interestingly old scars have been unaffected. He recently developed stiffness of his feet but plantar fibromatosis was not clinically evident.

Investigations: Full blood count, ESR, serum urea and electrolytes, proteins, renal and liver function tests and muscle enzymes were normal. Pulmonary function and serum angiotensin-converting enzyme were also normal. Antinuclear and anticytoplasmic antibodies were negative.

X-rays showed a bilateral punched-out erosive peripheral arthropathy of the hands, feet and shoulders. There was no soft tissue calcification. A bone biopsy from a finger showed increased bone resorption with many osteoclasts eroding bone trabeculae, associated with clusters of macrophages and osteoblasts.

Surgical biopsies of several cutaneous nodules showed similar changes. Extensive interwoven bundles of collagen were seen occupying the whole dermis at scanning magnification. On higher power, cellular proliferation was evident between the broad bundles (Figure 3). There was also extensive mucin deposition between the bundles on one biopsy. The fibroblasts were monomorphic but with many

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