CASE REPORT

Pseudocyst formation in retroperitoneal fibrosis relapse

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ABSTRACT. We describe a 45-year-old male patient with recurrent retroperitoneal fibrosis (RPF), in whom a pseudocyst in the peri-aortic fibrotic mantle was diagnosed. Without any intervention other than oral treatment with tamoxifen, the pseudocyst showed significant regression. Although rare, pseudocyst formation may sometimes appear in RPF and may mimic other benign and malignant conditions. This is the first paper to describe pseudocyst formation in an RPF relapse.

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Retroperitoneal fibrosis (RPF) is a rare collagen vascular disease with a chronic relapsing course. RPF is characterised by a non-specific chronic inflammation, usually surrounding the abdominal aorta and iliac arteries. It may extend into the retroperitoneum, thereby entrapping and possibly obstructing surrounding structures, most notably the ureters. Although usually idiopathic, RPF can also be secondary to the use of certain drugs, malignancies, infections or surgery. The aetiology of idiopathic RPF (IRPF) is unclear. A possible autoimmune aetiology has been postulated in addition to the concept that the disease is a result of an inflammatory response to antigens in the atherosclerotic plaques of the abdominal aorta [1]. Immunosuppressive and/or hormonal therapy has now replaced surgical treatment as the first treatment of choice [2]. CT and MRI are preferred for the diagnosis of iRPF. Both can provide a virtually characteristic picture, thereby obviating the need for biopsy. Furthermore, nuclear imaging with ⁶⁷Ga SPECT or ¹⁸F-FDG PET can be a useful tool in assessing disease activity, thus indicating a potential response to medical treatment [3,4]. (Pseudo)cyst formation in areas of iRPF is an atypical appearance and has been reported in only two cases, by Feldberg and van Waes in 1987 [5]. We present a patient who developed a retroperitoneal pseudocystic mass four years after initial RPF diagnosis. This is the first time pseudocyst formation has been described in a relapse of RPF.

Case report

In 1998, a 39-year-old male patient presented with unspecific abdominal complaints. Relevant laboratory parameters included a raised erythrocyte sedimentation rate (ESR) (24 mm/h) and C-reactive protein level

(13 mg/L). A CT scan revealed a homogeneous, well-circumscribed, slightly enhancing peri-aortic soft-tissue mass with a maximal anterior–posterior diameter of 15 mm. On the basis of these characteristic imaging findings and the absence of additional clues to suggest (occult) malignancy or infectious processes, a diagnosis of RPF was established. The patient was treated with tamoxifen 20 mg twice daily for a duration of two years. On this regimen, his complaints abated completely. Follow-up CT scans showed moderate regression and a residual peri-aortic mass one year after diagnosis.

In 2004, the patient presented with renewed complaints of malaise and lower back pain of four months in duration. CT scanning showed a novel, well-circumscribed, hypodense mass abutting the ventral RPF mantle in conjunction with a further overall regression of the initial peri-aortic mass (Figure 1). The MRI that was subsequently performed to better delineate the abnormal mass in relation to vascular structures confirmed an apparently pseudocystic mass of 30 × 23 mm (Figures 2 and 3).

Although atypical in its radiological presentation, the mass was interpreted as a relapse of RPF and tamoxifen therapy was re-introduced. No attempt was made to puncture the pseudocystic mass. Within two weeks, the patient felt better and his lower back pain gradually diminished. Likewise, laboratory investigation showed normalisation of acute-phase reactants. Repeated gallium scintigraphy showed the disappearance of pathological gallium activity. Follow-up CT scanning during the next 3.5 years showed an ongoing, significant regression of the pseudocystic mass (Figure 4). There was no further regression of the residual/initial RPF mass during this period.

Discussion

Idiopathic retroperitoneal fibrosis has a virtual characteristic imaging pattern on CT and MRI. It presents as a solid, homogeneous mass mostly surrounding the

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Figure 1. Contrast-enhanced CT scan at diagnosis of iRPF relapse. A well-circumscribed hypodense mass with hyperdense, enhancing rim abuts the right ventral side of the subtle initial RPF mantle, just above aortic bifurcation. On the left of the pseudocystic mass, some of this initial typical periaortic RPF mass is seen surrounding an atherosclerotic abdominal aorta. A, abdominal aorta; c, pseudocyst; *, para-aortic IRPF mass.

abdominal aorta and iliac arteries, and it may envelope and possibly obstruct the ureters and/or inferior vena cava [1]. RPF can, however, have a very extensive localisation pattern and can even present as a solitary lesion, mimicking multiple other conditions. The iRPF mass is isodense to surrounding muscle tissue, and characteristically there is no displacement of great vessels or the psoas muscles. Encasement of the aorta, along with displacement of the aorta anteriorly and the ureters laterally, is suggestive of lymphoma or disseminated malignancy. On CT, contrast administration improves the visibility of the early, inflammatory stages of IRPF. The extent of contrast enhancement cannot,

Figure 2. T_1 weighted MR imaging after gadolinium shows a non-enhancing hypointense mass with rim enhancement. A, abdominal aorta.

Figure 3. T_2 weighted MR imaging shows a mass with high signal intensity consistent with fluid.

however, be used reliably to assess the metabolic activity of iRPF. On MRI, iRPF is hypointense on T_1 weighted images. On T_2 weighted images, its intensity is high in early active stages because of tissue oedema and hypercellularity, and low in the late fibrotic stages. Heterogeneous signal on T_2 weighted images suggests malignant retroperitoneal fibrosis, which is typically the result of breast, gastrointestinal or lung cancer [6].

To date, only two cases of pseudocyst formation associated with RPF have been described [5]. These pseudocysts were both in histologically proven RPF patients, one was identified at diagnosis, the other three months after presentation. The first patient underwent surgical removal of the cystic area. In the second patient, the cystic area subsided under prednisone treatment.

In light of this, we diagnosed this unusual cystic presentation of IRPF in our patient. Although no

Figure 4. Follow-up contrast-enhanced CT scan one year after re-introduction of tamoxifen treatment shows significant regression of the cystic RPF component. A, abdominal aorta.

histological proof was obtained, the patient's history of typical IRPF presentation on CT, his former good response to tamoxifen treatment and the regression of the cystic lesion after a second course of tamoxifen therapy makes the diagnosis of iRPF very likely.

There are, however, other differential diagnoses that may be considered. Sclerosing mesenteritis is another group of fibrosing disorders that can affect the retroperitoneum. This is a rare and complex inflammatory disorder of the mesentery often associated with IRPF [7]. Cystic components have been described in sclerosing mesenteritis and are claimed to be the result of lymphatic or venous obstruction and necrotic change [8]. Imaging findings that support a diagnosis of sclerosing mesenteritis are the distribution of cystic components along the mesentery and the presence of calcification, which have not been reported in IRPF to date.

Cysts in the retroperitoneum are rare. If not congenital in origin, they are most probably formed post-traumatically (due to the resorption of a haematoma) or post-inflammatory (usually in direct contact with the primary site, for example, in pancreatitis). Although the majority (70–80%) of retroperitoneal masses are malignant in behaviour, none of them (*e.g.*, lymphoma, metastatic lymphadenopathy, germ cell tumours or liposarcomas) are likely to develop cystic degeneration [9]. No clues for malignancy, such as additional tumour implants, invasive growth, lymphadenopathy or ascites, were found in our patient. Lymphangioma and peripheral nerve sheath tumours are benign neoplasms of the retroperitoneum that may undergo cystic degeneration. However, these either demonstrate differential imaging characteristics or occur in a different age group from that typical for IRPF.

It is difficult to ascertain whether the onset of pseudocyst formation is related directly to activity of IRPF disease. If we include the present case, the pseudocyst was present at the time of (recurrent) presentation of active iRPF disease in two of the three reported cases [5]. In addition, the pseudocystic area regressed in two patients following the initiation of medical treatment, concurrent with improvement of clinical and laboratory signs and CT-documented

regression of the fibrotic mass. Chemical analysis in the case that was subsequently treated surgically identified the cyst as chylous in origin [5]. In analogy to the proposed theory as to the possible origin of cyst formation in sclerosing mesenteritis [8], one might speculate that in active RPF disease, the vascularised and oedematous mass may result in lymphatic obstruction and the subsequent formation of a pseudocyst.

In conclusion, we describe a rare cystic presentation of a relapsing IRPF mass that responded well to tamoxifen therapy. Cystic masses in IRPF may be confused with a range of other conditions, both benign and malignant.

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