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Retroperitoneal fibrosis with pancreatic involvement – radiological appearance

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Summary

Background:

Retroperitoneal fibrosis or Ormond's disease is an uncommon process characterized by fibrous tissue proliferation in the retroperitoneum, usually involving the aorta, inferior vena cava and iliac vessels. Obstructive hydronephrosis is often observed due to ureteral entrapment. This report presents a case of the peripancreatic location of the disease. The role of CT and MRI in establishing diagnosis of retroperitoneal fibrosis in an atypical site is discussed.

Case Report:

A 52-year-old woman with Hashimoto's thyroiditis was admitted to hospital because of pain suggesting renal colic. The patient was subjected to ultrasound, CT, and MRI which did not confirm urolithiasis but revealed pancreatic infiltration. Partial pancreatectomy, left-sided adrenalectomy and splenectomy were performed. Retroperitoneal fibrosis was diagnosed in the histopathological examination. A few weeks after surgery, a complication such as pancreatitis developed. Repeat CT confirmed it and showed right hydronephrosis secondary to ureteral involvement by a mass adjacent to the common iliac artery (defined as a typical manifestation of retroperitoneal fibrosis). Nephrostomy and conservative treatment improved the clinical state of the patient. No progression of the process was observed in the follow-up examinations.

Conclusions:

Atypical retroperitoneal fibrosis remains a diagnostic challenge. Imaging techniques CT and MRI are useful tools for evaluating the extent of Ormond's disease. An unusual distribution of the process (e.g. peripancreatic location reported in this study) requires histopathological assessment to establish the final diagnosis.

Key words:

retroperitoneum • fibrosis • CT • MRI

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Background

Retroperitoneal fibrosis (RF) is a rare process characterized by extensive development of inflammatory fibrotic tissue in the retroperitoneum.

The process usually involves the aorta, inferior vena cava and iliac vessels. Hydronephrosis frequently occurs due to ureteral obliteration. Atypical locations of RF were reported, such as perinephric, peripancreatic, periduodenal and pelvic [1,2].

Primary (idiopathic) form of RF, also called Ormond's disease, accounts for about 70% of cases. Immunological etiology of RF is strongly suspected due to frequent association

of the condition with autoimmune disorders such as Hashimoto's thyroiditis, rheumatoid arthritis, lupus erythematosus, Wegener's granulomatosis and autoimmune pancreatitis [3–5].

Secondary RF can be a result of: aortic aneurysm, previous radiotherapy, abdominal operation, trauma, primary neoplasms (lymphomas, sarcomas) or metastatic tumors in the retroperitoneal space, infections (pyelonephritis, tuberculosis, histoplasmosis) and drugs (ergotamine, methyl dopa, some beta-blockers, bromocriptin).

Clinical symptoms of RF are non-specific. The most common are back pain due to ureteral obstruction and

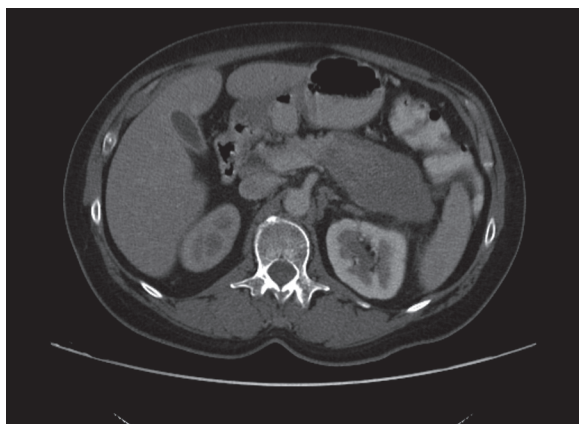


Figure 1. Enhanced CT: enlarged pancreatic tail appears as hypodense compared to normally-enhanced pancreatic body.

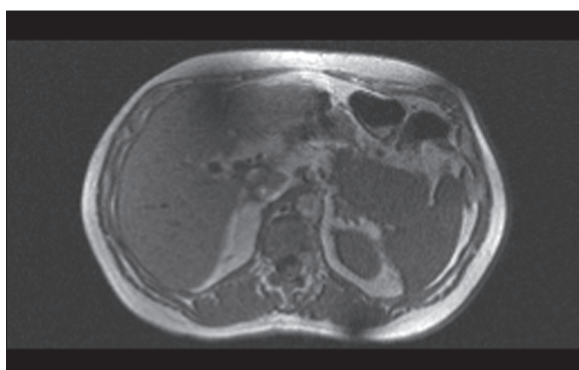


Figure 2. MRI: T1-weighted image revealed a significant enlargement of the pancreatic tail.

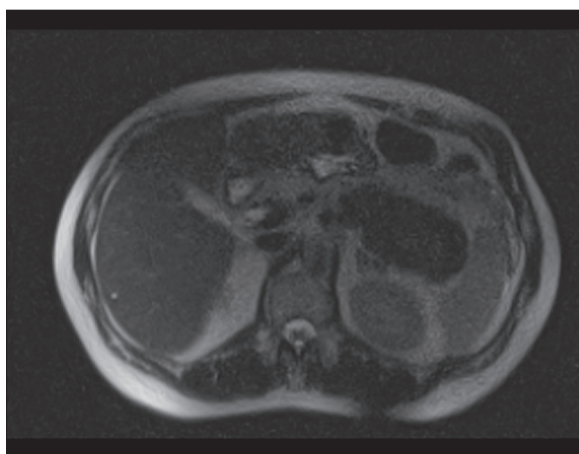


Figure 3. MRI: T2-weighted image showed an enlarged pancreatic tail with a low signal intensity, typical for the pancreatic parenchyma.

secondary hydronephrosis. Other clinical findings are anorexia, fatigue, anemia, weight loss and mild fever. In the late stage of the disease, progressive renal failure is noted [5]. Due to the multi-faceted nature of the Ormond's disease, the diagnosis relies heavily on radiological findings.

In the study, a case of peripancreatic RF distribution was presented and the value of imaging techniques for establishing the diagnosis was analyzed.

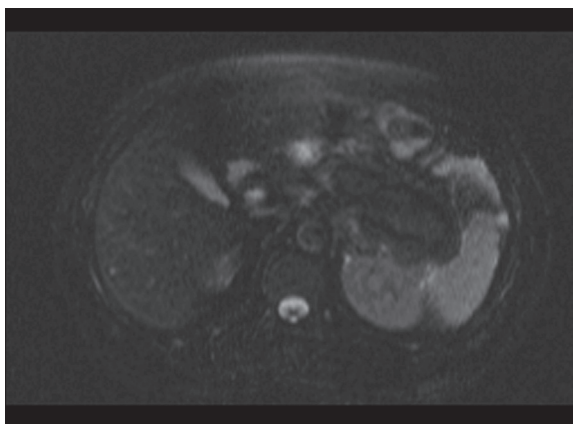


Figure 4. MRI: T2-weighted image in the sequence with fat suppression showed no hyperintense lesion in the region of the pancreatic tail.

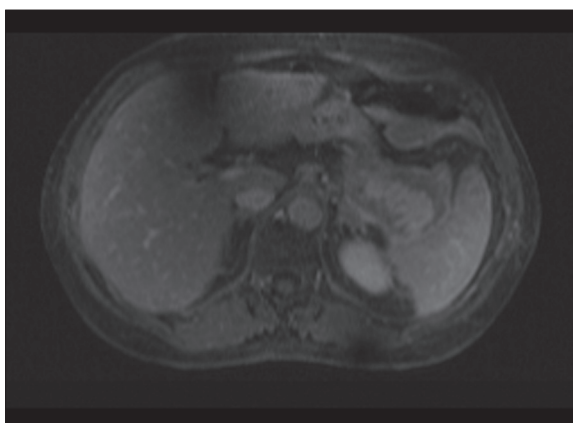


Figure 5. MRI: post-contrast T1-weighted image in the sequence with fat saturation demonstrated a well-delineated tissue around the pancreatic tail, less enhanced than the pancreatic parenchyma.

Case Report

A 52-year-old woman with a history of Hashimoto's thyroiditis was admitted to the Emergency Unit because of back pain suggesting renal colic. Her initial ultrasound did not confirm urolithiasis, but a mass in the left adrenal gland was suspected. The region of the pancreas was poorly visible on sonograms. The patient underwent further diagnostics in the Department of Endocrinology.

Computed tomography performed before and after contrast medium administration demonstrated hypodense infiltration of the pancreatic tail and body penetrating towards the splenic hilum and adrenal gland (Figure 1).

MR imaging depicted enlarged pancreatic tail with an adjacent part of the pancreatic body, which appeared as isointense with the pancreatic head on T1- and T2-weighted images (Figures 2, 3). The sequence with fat suppression based on T2-weighted images did not show any hyperintense lesions (Figure 4). Post-contrast MR scans, including sequence with fat saturation, revealed a well-delineated tissue around the pancreatic body and tail which enhanced less intensively than the pancreatic parenchyma (Figure 5).

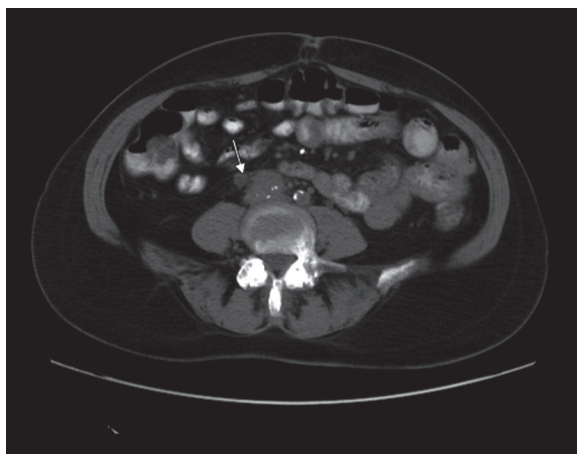


Figure 6. Postoperative CT: a soft-tissue mass surrounding the right common iliac artery (with calcifications in the wall of the vessel); dilated ureter next to the mass (arrow).

Neither the spleen nor the adrenal gland was involved. MR findings suggested a benign nature of the process.

Surgery was performed. Partial pancreatectomy and left-sided adrenalectomy were carried out with splenectomy due to splenic rupture during the operation.

Histopathological examination established the diagnosis of RF. A few weeks after surgery, pancreatitis developed. The complication was monitored with CT. The examination additionally revealed right hydronephrosis as a result of ureteral obliteration caused by the mass around the right iliac artery (Figure 6). The mass was defined as a typical manifestation of RF.

Nephrostomy and conservative treatment resulted in remission of hydronephrosis and pancreatitis. Administration of the steroids inhibited the expansion of the fibrous tissue.

The patient remains under regular care of the Outpatient Clinic of Endocrinology.

Discussion

Retroperitoneal fibrosis was first described in 1905 by Albarran. In 1948 Ormond reported this entity in the English literature [6,7].

Primary RF (idiopathic form), also termed Ormond's disease, is a relatively rare entity, with an estimated incidence of 1/200 000 population [8]. It is characterized by fibrous tissue proliferation along the posterior aspect of the retroperitoneal cavity. A few stages of RF have been described, ranging from active inflammation to fibrous scarring [9]. The idiopathic RF can be established by exclusion of neoplastic processes, infections or hematomas.

Non-specific clinical symptoms, such as poorly localized back pain, general malaise, weight loss, anemia, sometimes mild fever, features of renal failure lead to a situation in which the patients with RF are diagnosed at various clinics (departments of internal medicine, urology, nephrology,

surgery, oncology). In the described case, the patient was admitted to the Department of Endocrinology because of an adrenal mass found on ultrasound examination. She presented uncharacteristic back pain.

Worth emphasizing is the fact that the patient had a history of Hashimoto's thyroiditis. Association of RF with Hashimoto's thyroiditis as well as Riedel's thyroiditis and Graves' disease was reported by Pizzini, Armigliato and others [10–13]. Both the evidence of a correlation between RF and fibrosing lesions (e.g. sclerosing cholangitis) or autoimmune disorders, as well as RF response to immunosuppressive therapy support the opinion that RF is a manifestation of a systemic immunological disorder. According to one of the theories, the relationship between RF and systemic fibrosis can be explained by the presence of ceroid in the atheromatous plaque of blood vessels that may act as an antigen and provoke an immune response [1,14].

A typical distribution of RF includes the region around the abdominal aorta and inferior vena cava with spreading from the level of the renal hila into the pelvis along the iliac vessels. There is no elevation of the aorta from the spine in the typical picture of RF.

In several studies, an unusual location of RF has been described. Some authors reported perirenal involvement [15–17]. Isolated perinephric distribution of RF requires differentiation from lymphoma, histiocytosis, amyloidosis and extramedullary hematopoiesis.

Vivas et al. [1] reviewed 30 patients with Ormond's disease; among them, there were 12 persons with RF in atypical sites such as: pelvic (7 cases), peripancreatic (4), periduodenal (1).

In our study, RF manifestation first discovered in radiological examinations mimicked pancreatic infiltration. There is little literature data describing pancreatic involvement in Ormond's disease [18–23]. Radiological findings in the analyzed studies included tumor-like formations (more frequent in the pancreatic head) or diffuse pancreatic/peripancreatic infiltrations (more often in the pancreatic body and tail).

In our material, CT findings showed diffuse infiltration of the pancreatic tail and body, which was hypodense compared to the pancreatic head on pre-contrast CT scans. Contrast enhancement of the lesion was less intense than in the non-affected part of the pancreas.

MR findings seem to be more precise in the determination of the lesion benignity. Several reports have emphasized the value of T2-weighted images in differentiation between the early and the chronic phase of RF [1,15,17,24]. The features of the late stage of RF observed in T2-weighted images suggest a benign process whereas the active phase may mimic malignancy in some cases. In T2-weighted images, mature fibrous tissue (composed predominantly of collagen) appears as hypointense in contrast to the early fibrotic formation which shows a high, heterogeneous signal intensity. This is a result of abundant fluid content and hypercellularity in acute fibrosis. Contrast enhancement of

RF may appear after several months from onset although it decreases gradually. In the described case, T2-weighted images revealed a low signal intensity in the lesion (including the sequence with fat suppression). Post-contrast MR scans showed that the tissue was located in the peripancreatic region. Intermediate contrast enhancement of the lesion was seen, especially in the sequence with fat saturation.

A follow-up CT depicted another focus of RF, which was located typically around the common iliac artery.

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Conclusions

In conclusion, RF may simulate neoplasm, especially if the disease is located in an unusual site. Both CT and MRI enable evaluation of the extent of the process. MR imaging is superior to CT in identifying the chronic fibrotic tissue due to a low signal intensity of mature fibrosis on T2-weighted images. Nevertheless, the final diagnosis requires histopathological verification.