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Case Report

Systemic Vasculitis Mimics Pancreatic Tumor

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ABSTRACT

The systemic vasculitides are heterogeneous conditions of unknown etiology characterized by inflammation and necrosis of different sized blood vessels. The epidemiology of the systemic vasculitides is poorly documented, but it is a rare disease. The abdominal manifestation of systemic vasculitis (SV) is a rarity. A 67-year-old man with a history of diabetes insipidus and bilateral ear canal stenosis presented with bilateral lower abdominal pain for three days duration and postprandial nausea and regurgitation for two weeks duration. He also complained of weight loss during the last year, decrease in general performance, and impaired hearing associated with ear bleeding. The patient was diagnosed with unspecified acoustic meatus stenosis by ear, nose, and throat (ENT) specialist. Computed tomography (CT) scan revealed a pancreatic lesion, which was biopsied under endosonographic guidance without any informative result. For further histological evaluation laparotomy was indicated and multiple histological materials were taken. Unfortunately, histological results revealed no specific diagnosis either. In the blood tests there were no signs of autoimmune or rheumatologic disease. By the daily deterioration of the patient's general condition, we carried out a Positron emission tomography-CT scan, which revealed SV. Under cortisone therapy the patient's complaints obviously declined, the patient rejected further investigation such as vascular biopsy. The intraabdominal vasculitis represents an unusual pathological finding, which, however, manifests itself only as a rare variation of an everyday medical constellation. The case impressively illustrates how difficult is to establish the correct diagnosis of abdominal vasculitis.

Case Report

In January 2019, a 67-year-old man presented with lower abdominal pain accompanied by postprandial vomiting and regurgitation. He suffered from weight loss since one year, accompanied by hearing problems and bleeding of both ears. An ear, nose, and throat ENT-specialist had already seen the patient, diagnosing unclear stenosis of the ear canals on both sides. Physical examination showed tenderness in the whole abdomen without signs of peritonismus. Laboratory results showed leukocytosis and elevation of C - reactive protein (CRP) (Figure 1). Other laboratory results included lipase, amylase, renal tests, transaminases, and bilirubin with normal values.

Abdominal CT revealed a pancreatic body mass with infiltration of the mesenteric root and narrowing the superior mesenteric artery (SMA) (Figure 2). Based on these CT scans an inflammatory of the pancreatic tumor was suspected, and an antibiotic treatment with piperacillin/tazobactam was initiated, combined with administration of octreotide. In 2017 the patient was diagnosed with central diabetes insipidus due to enlargement of the pituitary gland and there treated with desmopressin. Further co-existing diseases included ear canal stenosis with newly diagnosed hypacusia and otorrhagia, arterial hypertension, coronary heart disease with multiple stent implantation und concomitant chronic heart failure. An endosonography-guided transgastric biopsy was taken. Histologic examination showed slight histiocytosis without any malignant criteria. The gastroduodenoscopy itself showed no abnormalities. Autoimmune pancreatitis and infectious hepatitis could be excluded on the basis of laboratory screening.

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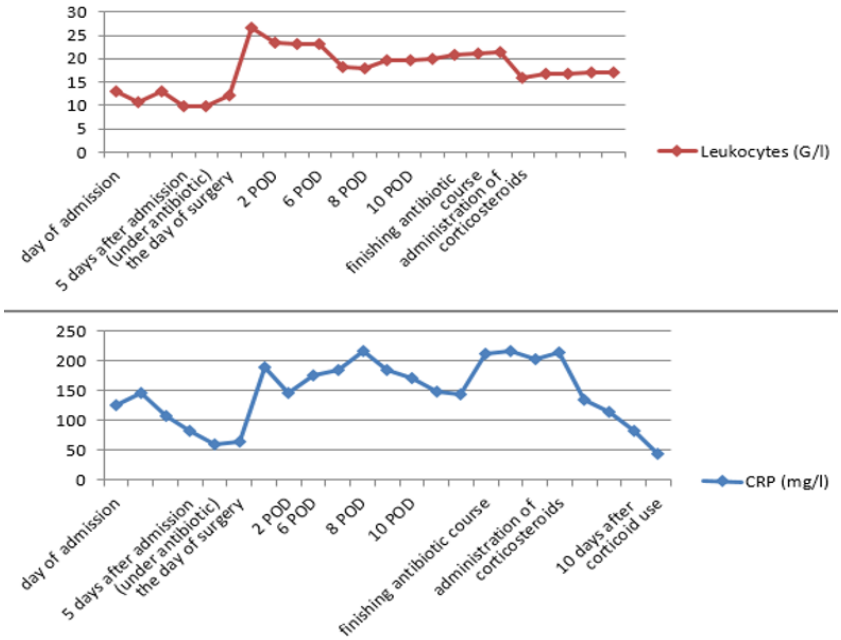

Figure 1: Laboratory overview.

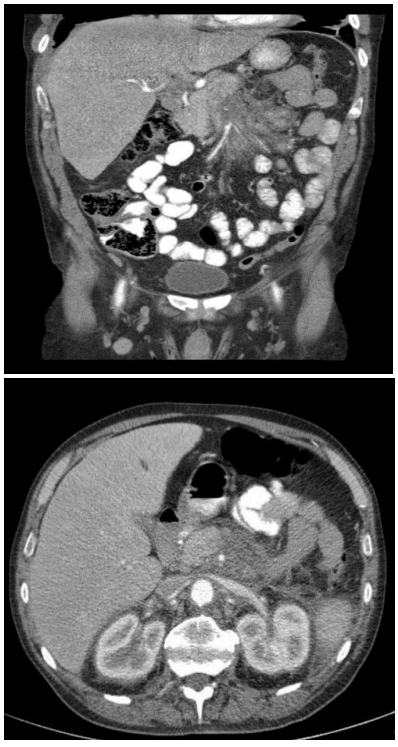

Figure 2: Abdominal CT, on the day of admission.

Within five days of antibiotic treatment showed the patient no clinical improvement. Control CT scan confirmed no change in imaging. Therefore, a laparotomy with exploration of the pancreas and multiple biopsies was performed. Intraoperatively, a big nodular tumor was seen with sufficient blood flow in the superior mesenteric artery and unremarkable intestinal loops. The gallbladder showed wall thickening resulting in intraoperative cholecystectomy. In all biopsies of parapancreatic tissue a pronounced scarred fibrosis with chronic unspecific inflammation without malignancy was found. Postoperatively, there was a constant increase in CRP as well as increased dyspepsia with regurgitation and nausea. A CT abdomen was performed again, with no indication of any inflammatory focus except for the previously known unclear peripancreatic process. By the evaluation of urine status, chest X-ray, and echocardiography no focus of infection could be found. Despite continued antibiotic therapy, no improvement was observed. An oral administration of Nystatin was initiated after endoscopic detection of mycotic esophagitis. Despite this, there was no improvement in the dyspepsia symptoms.

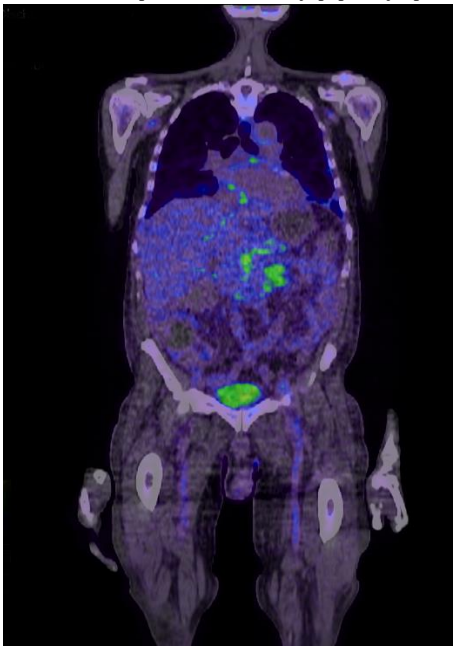

Figure 3: PET-CT - Arteriitis of aorta ascendens, also subclavian and femoral arteries.

The repeated blood culture test, after antibiotic therapy was terminated, resulted in neither bacteremia nor fungaemia. The procalcitonin level

was normal. In routine laboratory controls, a steady increase in CRP and leukocytes was observed (Figure 1). In addition, there was a significant increase in blood sedimentation rate. Comprehensive blood tests, including rheumatological diseases, revealed only an increase of the a1- and a2-microglobulins. PET/CT was initiated, as the described symptoms were not consistent with any pathology. Herein, the suspicion of systemic vasculitis (SV) was expressed with the involvement of the ascending aorta, and the subclavian and femoral arteries on both sides was suggested (Figure 3). A possible Ormond syndrome (OS) due to increased glucose metabolism in the retroperitoneum and unclear inguinal lymphadenopathies (IL) on both sides was described.

On the basis of these results, cortisone therapy was started. After that, the symptoms improved rapidly, and the inflammatory values also decreased rapidly. To clarify the IL, a tumor manifestation from the urogenital system was excluded. The patient was ophthalmologically examined. No organic disease could be detected. The temporal arteries were sonographically examined, and the suspicion of a halo sign was expressed on both sides. Due to a complete regulation of complaints under the established cortisone therapy, the patient did not wish any further diagnostic tests.

Discussion

SVs are mostly autoimmune diseases which, depending on the type, size, and localization of the affected vessels, offer a broad spectrum of manifestations from harmless, self-limiting to fulminant, life-threatening progressions. Vasculitis is classified according to the Chapel Hill Consensus Conference (CHCC) 2012 and most often types are: Giant cell arteritis (GCA), Takayasu arteritis (TA), polyarteritis nodosa (PN) and Kawasaki disease [1]. GCA is the most common systemic vasculitis in patients over 50 years of age (incidence 3.5 per 100,000). It affects cranial vessels, the aorta, and the arteries of the extremities. The typical symptoms of GCA are often misinterpreted and the urgently needed treatment delayed [2]. TA mainly affects younger women under the age of 40 [3].

The abdominal manifestation of SV is a rarity. There are few cases described in the literature where SV was the cause of abdominal pain [4, 5].The abdominal manifestation of vasculitis usually occurs as part of a systemic inflammatory process, especially in vasculitis of small and medium vessels, such as: PN and TA [6, 7]. Even rarer is the involvement of the gastrointestinal system in GCA [8, 9]. Retroperitoneal fibrosis (RF) is a rare inflammatory disease with a disease peak between the 5th and 6th decade of life. Men are affected about 3 times more frequently than women. Two-thirds of all RF are primarily idiopathic (= Ormond's disease). Approximately one-third of all RF is a secondary form (= Ormond syndrome) which can be triggered by inflammation, trauma, radiation, infection, malignancy, and medication or are associated with other fibrotic processes such as Riedel's goiter, sclerosing cholangitis, or mediastinal fibrosis [10].

The intraabdominal vasculitis represents an unusual pathological finding, which, however, manifests itself only as a rare variation of an everyday medical constellation. The case impressively illustrates how difficult is to establish the correct diagnosis of abdominal vasculitis. In our case, RF was most likely an OS in systemic arteritis, and not a coincidental finding of Ormond's disease. Even targeted biopsies from pathologically altered organs can be a false negative since the inflammatory vascular changes are mostly focal or segmental. This emphasizes the role of PET-CT in unclear abdominal findings.

Conflicts of Interest

None.

Disclosure

The work was conducted in University clinic of general, abdominal, and thoracic surgery, Hospital of Herford, Schwarzenmoorstr. 70, 32049, Herford, Germany.

Ethical Approval

The paper is exempt from ethical committee approval. The authors confirm that written informed consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Author Contributions

MK is the main author, who wrote the paper, CK and GW performed editing and literature search, and are senior authors. EK is the patient consultant and performed article correction.

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