RHEUMATOLOGY RESEARCH JOURNAL

Vol. 6, No. 1, Jan 2021,
Webpage: http://rheumres.org
Email:rheumres@gmail.com
ISSN:2476-5856
doi: 10.22631/rr.2021.69997.1115

Case Report Open Access

Extracranial as Silent Giant Cell Arteritis: Case Report

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GCA (Giant cell arteritis) is a granulomatous vasculitis of large arteries. Frequently typical cranial symptoms are observed, but sometimes nonspecific extracranial involvements are dominant. Diagnosis of this “occult” or “extracranial” GCA as a medical emergency is crucial to preventing irreversible complications. The current article presents the case of a 52-year-old man with no cranial manifestations who developed acute peritonitis and died. Elevated inflammatory markers without cranial manifestations should cause extracranial GCA to be considered. Delayed diagnosis in GCA, especially the extracranial type, could lead to severe, irreversible complications.

Keywords: Case report, Extracranial, Giant cell arteritis

Introduction

Giant cell arteritis (GCA) is a granulomatous vasculitis of medium and large arteries, frequently presenting with typical cranial symptoms, but sometimes nonspecific extracranial involvements are dominant. Diagnosis of this “occult” or “extracranial” GCA as a medical emergency is crucial to preventing irreversible complications [1, 2]. GCA vasculitis involves large- to medium-sized arteries, especially the aorta and proximal branches. Its classic presentations as a cranial arteritis comprise temporal headache, visual disturbance, and jaw claudication [2, 3]. In recent years, interest in attaining knowledge about cases with non-classic manifestations and involvement of extracranial arteries has increased [1]. Because of the non-specific manifestations of extracranial GCA, it seems that actual incidence prevalence, which is reported as only 1.6-32.8 per 100,000 people, has been underestimated [1]. Common symptoms of extracranial GCA include fever, anorexia, malaise, weight loss, polymyalgia, and muscle weakness. Some vascular manifestations are more specific but less common, such as Raynaud’s phenomenon, digital ischemia, decreased pulse, limb claudication, arterial bruits, and signs of cerebral ischemia. In comparison with cranial GCA, extracranial GCA is more prevalent in women, and the onset of disease occurs at younger ages. Moreover, there is a longer delay in diagnosis. Therefore, in patients over 50 years of age with constitutional raised inflammatory markers, a diagnosis of extracranial GCA must be considered [1]. High risk for developing aneurysms and dissection of the aorta (especially thoracic segment) necessitates prompt diagnosis and treatment [3].

Because of the non-specific signs, symptoms, and blood tests and the difficulties in biopsying affected arteries, extracranial GCA is often diagnosed based on imaging. Acute aortic pathology as the first clinical presentation with a high mortality rate (44-80%) raises suspicion of previously established extracranial involvement. The gold standard for diagnosing extracranial GCA used to be conventional angiography, but that has been replaced with non-invasive methods [3]. Despite the lack of a specific laboratory marker, acute phase reactants (platelets, erythrocyte sedimentation rate, and/or C reactive protein) are raised in most patients. Normal values, however, in the presence of strong clinical suspicion would not rule out the diagnosis [3]. In patients with typical cranial presentation, duplex ultrasonography of the temporal artery is a proper diagnostic modality. In addition, diagnosing patients with predominant extracranial manifestations and atypical ones may be aided by other imaging techniques, such as CT (computed tomography), MRI (magnetic resonance imaging), CT-angiography, and PET (positron emission tomography). The widespread use of these new imaging techniques may lead to the identification of clinically silent large vessel involvement, and this will probably raise the rate of incidence [3].

In diagnosing GCA, a specialist clinical evaluation, which can be based on signs, symptoms, and laboratory

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Received: 23 February 2020; **Accepted:** 13 July 2020

findings, is more important than other factors. Furthermore, this disease is considered a medical emergency. Therefore, frontline clinicians, such as general practitioners and clinicians working in acute care, should have information on the management and initial treatment with high-dose corticosteroids and the conditions for referring patients to specialized centers [4]. High doses of corticosteroids used to treat GCA should be reduced to lower doses during the course of making a definitive diagnosis. Clinical evidence suggests initial glucocorticoid therapy for GCA in patients without ischemic visual loss, stroke, or tissue necrosis. It seems most patients respond symptomatically within 1–7 days to a 40–60 mg daily dose of prednisolone [4].

Case presentation

The patient in this report was a 52-year-old man with a history of inhaled opium addiction for 20 years. The symptoms were abdominal pain (colic and peri-umbilical pain; exacerbation with eating) and diarrhea (watery stool without hematochezia associated with abdominal pain) in the prior 3-4 months. Abdominal peri-umbilical pain (suspicious of lead poisoning; treatment was initiated, and partial improvement was observed) had begun 2 years prior to this study. After discontinuing opium use 3-4 months before the study, the patient's abdominal pain had become severe and intolerable. Meanwhile, diarrhea had begun 2-3 months ago. The patient had a history of appendectomy 4 years prior to the study. Generalized tenderness without rebound tenderness was observed in the abdomen. Acute vision loss in the right eye and hypertensive crisis began after 4-6 months previously. In a clinical examination, central retinal vein occlusion (CRAO) was observed. The requested laboratory tests indicated normal CBC but elevated ESR. Normal ANA, P-ANCA, Anti ds DNA and anticardiolipin IgG/IgM, β-2Glyco protein Ab IgG/IgM levels were observed. A colonoscopy revealed colon polyps and terminal ileum congestion, and antral ulcers and duodenal nodularity, patulous LES (lower esophageal sphincter), and sliding hiatal hernia (H.H) were observed in the endoscopy. According to the CT scan with dye results, scattered arteriosclerosis was observed. An enlarged aneurism 41 mm in diameter was seen in the proximity of the abdominal aorta, and the increasing diameter of the aorta in this part was also seen (Figure1). Obstructions in the celiac trunk artery, superior mesenteric artery, and right renal artery were observed in the CT scan (Figure 2). Several collateral arteries were replaced with mentioned branches. The hypertrophic inferior mesenteric artery and its collaterals were in charge of intestinal vascular nutrition. The diameter of the aorta was normal in other parts (Figure 3). There was no clue of dissection, aneurism in the aorta, nor liquid accumulation around it. The CT scan proposed inflammation of vasculitis. Treatment was initiated with prednisolone 1 mg/kg, ciprofloxacin and metronidazole, but during the treatment the patient's abdominal distension was exacerbated. In radiography, signs of peritonitis with free air intra-abdominally were observed. The disease course lasted 40 days. Contrary to treatments, the patient's condition led to acute abdomen, and eventually the patient expired.

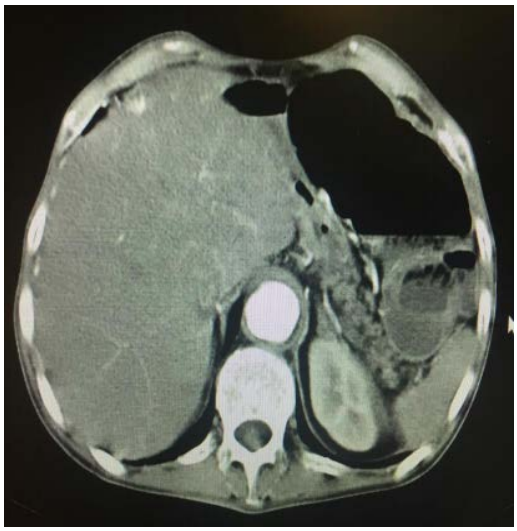

Figure 1. Increasing Diameter of Abdominal Aorta

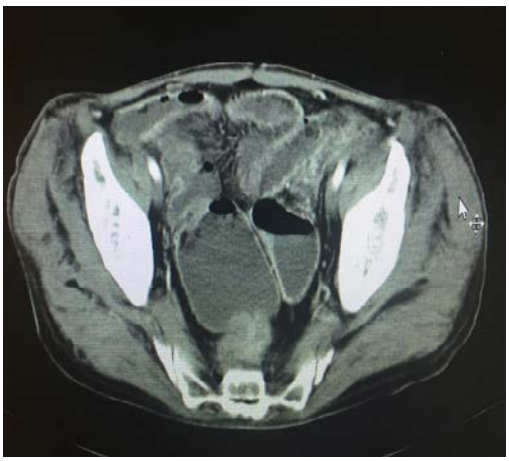

Figure 2. Obstruction of pelvic aorta trunk and branches

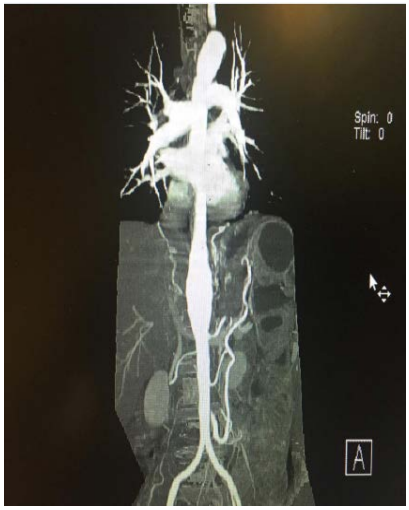

Figure 3. Spiral Angiography of Abdominal Aorta

Discussion

This paper considered a middle-aged man with a history of abdominal pain from 2 years previous to the study, which was exacerbated in the last 3-4 months. Laboratory tests have reported increasing ESR. Ultrasonography and spiral CT angiography revealed increasing diameters of aortas, obstruction of celiac trunk artery, superior mesenteric artery, and right renal artery. Several collateral arteries were replaced with the mentioned branches. Any hypertrophic inferior mesenteric artery and its collaterals was in charge of intestinal vascular nutrition. The diameter of the aorta was normal in other parts. Prednisolone, metronidazole, and ciprofloxacin were administrated, but abdominal distension was exacerbated, and in the radiography signs of peritonitis with free air, intra-abdominal was observed. The course of the disease led to acute abdomen and the eventual death of the patient.

Sakashita et al. (2021) published a case report about the autopsy of an 87-year-old Japanese woman with aortic dissection. She had a history of abdominal aortic aneurysm and endovascular aneurysm repair (EVAR) about two years prior to this study. Despite the absence of any characteristic symptoms of GCA, autopsy represented evident granulomatous inflammation in the dissected area and coronary arteries. Active arteritis was obvious in the upper and lower extremities. Eventually, seven days after admission, the patient died. In this case report, researchers suggest that the patient's manifestations could be the onset of GCA [5].

Large artery involvement in GCA is doubtful, but clinical features are estimated to occur in 9%–14% of cases accompanied by aortic aneurysm with or without dissection. Occult GCA with no clinical manifestations has been reported in almost 50% of cases before aortic dissection [6], which can explain the presentation and considered diagnosis of Sakashita et al.

Takase et al. (2019) presented a 76-year-old woman admitted to hospital for a high C-reactive protein (CRP) level but no remarkable findings on her thoracico-abdominal CT and head MRI. Administration of non-steroidal anti-inflammatory drugs (NSAIDs)caused a gradual decrease in CRP serum levels and improved the patient’s fatigue. Suddenly, on the 46th day after her first visit, the patient collapsed, cardiopulmonary arrest occurred, and she died. On pathological autopsy, the only cause of death found waws acute aortic dissection (Stanford type A) due to ascending aorta giant cell arteritis. In the histologic assessment, granulomatous vasculitis with giant cells was identified in the ascending aorta, thoracic descending, and abdominal aorta and their branches. Likewise, in the medium and small vessels of other plural organs, including the heart, liver, uterine corpus and its appendages, similar granulomatous vasculitis was also found. Therefore, a follow-up examination for CRP elevation could be beneficial in GCA as a more sensitive inflammatory reaction indicator [7].

This case was similar to the one in this article in the involvement of the aorta and its branch arteries. The only symptom of their patient was fatigue, which has been mentioned above as a common characteristic of extracranial GCA. Involvement of the heart or plural organs could explain the high percentage of fatalities in these patients without remarkable manifestations of GCA. Also, elevated inflammatory markers such as CRP in the case of Takase et al. and ESR in ours seem similar.

One of the important aspects of the current case was the rarity of the incidence of symptoms in these patients, especially abdominal pain and other gastrointestinal complaints. As far as we have searched, no similar case has been reported. Additionally, most of the cases, which comprise only a handful, have been in patients whose autopsy have determined GCA; therefore, it was not possible to evaluate and rule out the occurrence of this disease during the patient's lifetime.

Conclusion

Elevated inflammatory markers without cranial manifestations should cause consideration to be given to extracranial GCA. Delay of diagnosis in GCA, especially the extracranial type, could lead to severe, irreversible complications.

Acknowledgments

None.

Conflict of interest

None.

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