CASE REPORT

Temporal Arteritis Presenting with Gastrointestinal Symptoms in a Middle Aged Man

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Giant cell arteritis, also known as temporal arteritis, is a vasculitis of unknown etiology that classically involves the wall of the large to medium size. We are reporting a case of a young onset temporal arteritis presenting with gastrointestinal symptoms. The patient was a 48-year-old male who presented with a 2-week history of fever, diffuse abdominal pain, and malaise. He underwent a laparoscopic cholecystectomy after findings of elevated bilirubin and alkaline phosphatase as well as suspicion of porcelain gallbladder on ultrasound (or computed tomography scan). The patient subsequently developed painless, intermittent vision loss and unilateral headaches. A work-up included temporal artery biopsy, which showed marked lymphocytic infiltrate in the arterial wall consistent with temporal arteritis. The presentation of temporal arteritis may be atypical. We are reporting a case of temporal arteritis at a young age presenting mainly with gastrointestinal symptoms.

KEY WORDS: temporal arteritis; giant cell arteritis; vasculitis.

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Giant cell arteritis (GCA), also known as temporal arteritis, is a vasculitis of unknown etiology that classically involves the wall of the large to medium size arteries and only occasionally the small arteries. It is more prevalent at older age and usually presents with chronic headache, vision changes, myalgias, or fever of unknown origin. Disease onset before the sixth decade is considered exceptionally rare. We are reporting a case of temporal arteritis at a young age presenting mainly with gastrointestinal symptoms.

The patient was a 48-year-old Caucasian male with no significant past medical history who presented to an outside hospital with fever, lower back pain and abdominal pain, myalgias, mild nausea, and anorexia. He was a nonsmoker and did not use alcohol regularly. On examination he was in no acute distress but had a temperature of 38.4°C. His heart and lung examinations were normal. His abdomen was soft, diffusely tender, but without guarding. His initial blood work was unremarkable except for a hemoglobin of 10.6 g/dL (normal >14.0), elevated bilirubin 1.9 mg/dL (<0.2), and alkaline phosphatase 415 U/L (<126). Ultrasound and computed tomography (CT) scan of his abdomen showed calcification of the gallbladder wall (porcelain gallbladder) but no evidence of acute cholecystitis. However, based on these symptoms and the laboratory findings, a decision was made to pursue with laparoscopic cholecystectomy. Postoperative examination of

the gallbladder revealed only chronic cholecystitis with intramural calcification. Patient's symptoms improved briefly and he was discharged to home within a couple of days.

He returned to the emergency department at the same hospital the following day with increasing right upper quadrant abdominal pain and fever. His examination was remarkable for tenderness and guarding in right upper quadrant, which were felt to be due to his recent surgery. Except for transaminases, liver tests were still abnormal and the hepatitis A, B, and C serologies were negative. A repeat CT scan of abdomen was also unremarkable. His hepatobiliary iminodiacetic acid (HIDA) scan showed no evidence of biliary leak. He was treated with supportive therapy including analgesics.

The following day the patient complained of painless, intermittent vision loss and unilateral headaches. At that time, a request was made to transfer the patient to our institution. Vital signs upon arrival included a temperature of 37.3°C, heart rate of 94 per minute and blood pressure of 160/49. He was found to have a transient afferent pupillary defect in the right eye. His fundoscopic examination was normal. Temporal arteries were easily palpable, firm, but nontender and with normal pulses bilaterally. The abdomen had recent surgical scars, and was diffusely tender to palpation but no organomegaly was felt. The remainder of the physical examination was unremarkable.

Laboratory tests showed a white blood cell (WBC) count of $13.3 \times 10^3/\text{mm}^3$ (<10) with normal differential, hemoglobin 11.7 g/dL (>13.5), platelets of $894 \times 10^3/\text{mm}^3$ (<450). His basic metabolic profile, including liver studies, was normal except for an elevated alkaline phosphatase of 457 U/L (normal <125). The elevated sedimentation rate (ESR) was 140 mm/h (0 to 15) and C-reactive protein (CRP) was 23.6 mg/dL (0 to 1). The head magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) were unremarkable. A diagnosis of GCA was entertained based on his presentation and the patient was empirically started on high-dose systemic steroids. He became asymptomatic 48 hours later. Although the patient was initially reluctant to have a temporal artery biopsy, after consultation with ophthalmology, neurology, and rheumatology services, he eventually accepted, and this was performed 7 days after starting the corticosteroids. The biopsy showed marked mononuclear inflammatory infiltrate in the media of the arterial wall consistent with tem-

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FIGURE 1. Temporal artery biopsy showing mononuclear inflammatory infiltrate in the media of the arterial wall.

poral arteritis. Multinucleate giant cells were not identified (Fig. 1).

During the same hospitalization his platelet count rose up to $1039 \times 10^3/\text{mm}^3$. He underwent a bone marrow biopsy, which showed a normocellular marrow with increased myelopoiesis and megakaryopoiesis and no definitive evidence of myeloproliferative disorder. His hemogram gradually normalized after a few weeks of steroid therapy and the thrombocytosis was felt to be reactive to his systemic inflammatory process. On his last follow-up in the local clinic the patient was asymptomatic.

DISCUSSION

Giant cell arteritis is one of the most frequent vasculitis syndromes in western countries with an annual incidence around 28 per 1,00,000 in the population over the age of 50. The incidence increases with age and peaks in the seventh and eighth decade, while people less than 50 years old are generally felt to be spared.¹⁻³ The usual presentation of temporal arteritis includes headache, jaw claudication, visual loss, polymyalgia rheumatica, and fever.⁴ The American College of Rheumatology (ACR) criteria for GCA include: age >50, new onset headache, temporal arteries abnormalities on physical exam or pathology and elevated ESR. The combination of 3 out of these 5 criteria has a sensitivity of 93.5% and a specificity of 91.2%.⁵

Our patient's presentation was rather unusual because of his young age (less than 50) and atypical symptoms. In a population study from Olmsted county, Minnesota, over a period of 42 years none of the patients diagnosed with GCA were less than 50 years old.⁶ Conversely, 1 case series from France reported 3 patients with GCA who were less than 40 years of age.⁷ The reason for this age effect remains elusive.

The patient's initial symptom presentation was misleading. Aside from his fever and malaise, he had primarily gastrointestinal complaints, with abdominal pain, nausea, and anorexia. In combination with an elevated bilirubin and alkaline phosphatase, the presentation raised the suspicion of a hepato-biliary process. In fact, the patient underwent a prompt cholecystectomy only to find out that he had chronic cholecystitis with porcelain gallbladder. However, it is known

that porcelain gallbladder is merely the presence of calcium deposits in the gallbladder wall and it is usually not associated with either abdominal symptoms or abnormal liver tests.⁸ Furthermore, his liver tests remained abnormal after the gallbladder surgery. Although, other atypical symptoms of GCA have been reported, such as respiratory, cardiovascular, or nervous system-related, the predominant gastrointestinal presentation in this patient is rather unique.⁹ Other studies have shown that up to 20% of patients with GCA may have asymptomatic elevation of liver tests, primarily alkaline phosphatase.¹⁰ The reasons behind these laboratory abnormalities are unclear but they may be due to coexistent granulomatous hepatitis or hepatic artery vasculitis.¹¹ A case of lobular hepatitis associated with cranial nerve involvement from GCA has also been described.¹² Conversely, gastrointestinal symptoms may be present in other types of vasculitides. Almost half of the patients with polyarteritis nodosa or microscopic polyangiitis can have gastrointestinal manifestations.¹³

This patient was difficult to diagnose initially because of his young age, lack of classical symptoms, as well as the presence of symptoms and signs suggestive of an alternative diagnosis. However the presence of persistent fever, myalgias, and elevated liver tests suggested a systemic inflammatory process, including vasculitis. This may be further supported by the lack of a clear-cut diagnosis to explain the hepatic abnormalities. A cholestatic picture may be the expression of an intra- or extrahepatic bile duct obstruction but our patient's imaging studies were unremarkable and his transaminases were normal. His symptoms were not suggestive of an acute biliary process, such as biliary colic or acute cholecystitis. Furthermore, his gallbladder only showed evidence of chronic cholecystitis and calcification on pathological examination, neither of which should cause this patient's presentation. Finally, the surgical intervention had provided the means for an intraoperative liver biopsy, had the diagnosis been in doubt at the time of the surgery. Although this may have hastened a final diagnosis of GCA, this opportunity was missed. The diagnosis became more apparent after the vision loss and the other inflammatory markers were corroborated and the patient received therapy with high-dose corticosteroids with excellent results.

In the end, our patient met 4 of the proposed 5 ACR criteria for a definitive diagnosis of temporal arteritis. Thus, the association of headache, vision loss, elevated ESR, presence of infiltrates in tunica elastica of the temporal arteries, and prompt response to therapy with steroids leaves little doubt about the diagnosis even in the absence of classic giant cells in the pathology specimen.

One other interesting finding in this patient was the markedly elevated platelet count peaking at $1,039 \times 10^3/\text{mm}^3$. Although platelet levels can be elevated in any acute illness, a level over $1,000 \times 10^3/\text{mm}^3$ can be indicative of a myeloproliferative disorder. An association between temporal arteritis and myeloproliferative disorders including essential thrombocytosis has been previously reported.¹⁴ However, the bone marrow biopsy in this patient showed only reactive changes as mentioned above. Moreover, his hemogram returned to normal after a few weeks of antiinflammatory therapy.

In conclusion, we are presenting a case of early onset temporal arteritis presenting with gastrointestinal symptoms. Even if liver test abnormalities are not uncommon in patients with GCA, a predominantly gastrointestinal presentation is

unusual. The diagnosis was only made after the patient's symptoms failed to improve following a noncomplicated laparoscopic cholecystectomy. Therefore, we believe that temporal arteritis should be considered in the differential diagnosis of individuals who present with otherwise unexplained gastrointestinal and hepatic abnormalities especially when there are other symptoms or signs suggestive of a systemic inflammatory process. The recognition of nonclassical manifestations of GCA may lead to a more frequent and earlier diagnosis of the disease and, consequently, may prevent dangerous complications.

REFERENCES

1. **Bengtsson BA, Malmvall BE.** The epidemiology of giant cell arteritis including temporal arteritis and polymyalgia rheumatica. Incidences of different clinical presentations and eye complications. *Arthritis Rheum.* 1981;24:899-904.
2. **Huston KA, Hunder GG, Lie JT, Kennedy RH, Elveback LR.** Temporal arteritis: a 25-year epidemiologic, clinical, and pathologic study. *Ann Intern Med.* 1978;88:162-7.
3. **Salvarani C, Cantini F, Boiardi L, Hunder GG.** Polymyalgia rheumatica and giant cell arteritis. *N Engl J Med.* 2002;347:261-71.
4. **Smetana GW, Shmerling RH.** Does this patient have temporal arteritis? *JAMA.* 2002;287:92-101.
5. **Hunder GG, Bloch DA, Michel BA, et al.** The American college of rheumatology 1990 criteria for the classification of giant cell arteritis. *Arthritis Rheum.* 1990;33:1122-8.
6. **Salvarani C, Gabriel SE, O WM, Hunder GG.** The incidence of giant cell arteritis in Olmsted county, Minnesota: apparent fluctuations in a cyclic pattern. *Ann Intern Med.* 1995;123:192-4.
7. **Genereau T, Herson S, Piette JC, et al.** Temporal arteritis in young subjects. A trial of nosological classification apropos of 6 cases. *Ann Med Interne (Paris).* 1992;143:303-8.
8. **Horton DJ, Bilhartz LE.** Gallstone disease and its complications. In: Sleiseger and Fordtran. 7th edn. Saunders; 2002:1065-90.
9. **Hellman DB.** Temporal arteritis: a cough, toothache, and tongue infarction. *JAMA.* 2002;287:2996-3000.
10. **Ilan Y, Ben-Chetrit E.** Liver involvement in giant cell arteritis. *Clin Rheumatol.* 1993;12:219-22.
11. **Pedro-Botet J, Lopez MJ, Barranco C, Coll J, Bruguera M.** Granulomatous hepatitis and giant cell arteritis. *Am J Gastroenterol.* 1994;89:1898-9.
12. **Killer HE, Holtz DJ, Kaiser HJ, Laeng RH.** Diplopia, ptosis, and hepatitis as presenting signs and symptoms of giant cell arteritis. *Br J Ophthalmol.* 2000;84:1319-20.
13. **Kurland L, Chuang T, Hunder G.** The epidemiology of systemic arteritis. In: Lawrence RC, Shulman LE, eds. *Epidemiology of the Rheumatic Diseases.* New York: Gower; 1984:196-205.
14. **Espinosa G, Font J, Munoz-Rodriguez FJ, Cervera R, Ingelmo M.** Myelodysplastic and myeloproliferative syndromes associated with giant cell arteritis and polymyalgia rheumatica: a coincidental coexistence or a causal relationship? *Clin Rheumatol.* 2002;21:309-13.