Simultaneous Presentation of Crohn’s Disease and Takayasu Arteritis in a Teenage Patient

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Abstract
A 14-year-old female with no significant medical history presented with hypertensive urgency, in the setting of 4 to 6 weeks of diarrhea, abdominal pain, headaches, anemia, weight loss, and high blood pressures. Her evaluation revealed signs of a systemic inflammatory process that was most suspicious for inflammatory bowel disease. However, when her hypertension was evaluated with a renal Doppler ultrasound, there were signs of narrowing, stenosis, and hypoplasia that led to a diagnostic angiogram of the abdominal aorta. Full body positron emission tomography scan revealed multiple areas of stenosis and aortic thickening with enhancement compatible with Takayasu arteritis. She received prednisone, methotrexate, and infliximab with marked improvement in her clinical symptoms and inflammatory markers.

Keywords
rheumatology, radiology, imaging, pediatrics, cardiology

Introduction
Rheumatologic conditions and other autoimmune conditions may coexist, overlap, or precede one another in a single patient. For those who have unusual symptomatology that fails to align with a single disease process, the treatment team should broaden the differential diagnosis. Inflammatory bowel disease (IBD) can have extraintestinal manifestations involving any organ system. This can make it challenging to identify if a constellation of symptoms is a new primary disease or an IBD-associated process. Most case reports in the literature describe ulcerative colitis or Crohn’s disease preceding aortitis or Takayasu arteritis (TA) by months or years. There are very few case reports of the concurrent onset of both diseases in childhood. We discuss a case of an adolescent patient with multiorgan disease who presented with significant gastrointestinal (GI) and vascular disease simultaneously.

Case Presentation
Our 14-year-old female with no significant past medical history presented to the pediatric intensive care unit with headache and hypertensive urgency in the setting of 4 to 6 weeks of nonbloody diarrhea, abdominal pain, anemia, weight loss, and high blood pressures noted by her primary care physician. She also noted associated abdominal fullness, pain with eating, and early satiety. Social history included lack of sexual activity, smoking, or alcohol use. She was active in volleyball. There was no recent travel. Family history included Crohn’s disease in her father and her sister. Per the pediatrician’s workup prior to presentation, she had normal basic laboratory work, thyroid studies and electrocardiogram.

Physical Examination
She was emaciated, alert, afebrile, and hypertensive (blood pressure 180/100 mm Hg) on presentation. She had remarkable HEENT (head, ears, nose and throat), lung, and...
skin examinations. Her abdominal examination was notable for a mid-line bruit. Her cardiovascular examination revealed no murmurs, rubs, or gallops, normoactive precordium, normal, and symmetric pulses throughout. There were no neurologic deficits. Musculoskeletal examination was unremarkable.

**Diagnostics**

Given her hypertensive urgency on presentation, she had screening laboratory tests, a urinalysis, and a head computed tomography. Though slightly hyponatremic, basic metabolic panel was otherwise unremarkable. She had no leukocytosis. Urinalysis and head computed tomography were normal. Erythrocyte sedimentation rate and C-reactive protein were elevated at 78 mm/h and 9 mg/dL, respectively. Retroperitoneal ultrasound showed a small right kidney with diffuse cortical thinning. Echocardiogram had a mildly dilated ascending aorta. A renal ultrasound with Doppler was obtained and demonstrated blunted arterial waveforms consistent with stenosis of the main right renal artery.

**Clinical Course**

Given her multiorgan presentation, cardiology, nephrology, and gastroenterology were consulted. At that time, differential diagnoses included Crohn’s disease with extraintestinal manifestations, systemic lupus erythematosus, or systemic vasculitis. Her hypertension was thought to be secondary, not primary renal disease. Her aortic dilatation could have been caused by her anemia and acute weight loss. After initial stabilization of her hypertension during the pediatric intensive care unit stay, she was discharged. Her antinuclear antibody was negative and complements were normal. The following week, an angiogram was obtained to more fully investigate the right renal artery stenosis. This revealed multifocal areas of large vessel occlusion and stenoses (Figure 1A and B); angiogram findings were strongly suggestive of a systemic vasculitis. The patient had an esophagogastroduodenoscopy and colonoscopy, which showed chronic inflammatory changes on the esophagus and right colon with ulcerations, sparing the terminal ileum. The biopsies were consistent with Crohn’s disease. Her full body positron emission

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**Figure 1.** (A) Early phase aortogram in the anteroposterior view demonstrates no opacification of the celiac artery representing complete occlusion, very limited opacification of the right renal artery consistent with complete occlusion at the origin (white solid arrow), long segment stenosis of the superior mesenteric artery (white, dashed arrow), focal stenosis of the left main renal artery (asterisk), prominent inferior mesenteric artery with arc of Riolan variant (white, dotted arrow) providing retrograde flow to the superior mesenteric artery and the left main renal artery. Overall, the undulating caliber of the aorta is noted. (B) Late phase aortogram in the anteroposterior view demonstrates collateral arterial vessels around the right kidney providing minimal perfusion to parenchyma and retrograde filling of the right renal artery (white, solid arrow), improved opacification of the superior mesenteric artery with retrograde filling from the arc of Riolan variant (white, dashed arrow), and slight opacification of the common hepatic artery (black, solid arrow) likely due to retrograde filling of the celiac artery branches from an arc of Buehler variant.
tomography scan showed localized areas of aortic wall thickening with increased fluorodeoxyglucose uptake in multiple areas (Figure 2A) and cardiac magnetic resonance imaging (Figure 2B), concerning for TA. She was started on pulse dose steroids, infliximab, and methotrexate as induction therapy for her disease process. Her clinical symptoms have markedly improved and inflammatory markers have normalized on follow-up. Prednisone continues to be tapered and she remains on outpatient infliximab infusions as well as antihypertensive medications with improved blood pressure control.

Discussion
Takayasu’s arteritis is a rare condition, occurring in approximately 1 per million people per year in North America. The incidence of Crohn’s disease is about 100 per million per year. That makes TA coexistence with IBD even less likely.\textsuperscript{1,2} Cases of both diseases have been reported sporadically since the late 1970s.\textsuperscript{3} Interestingly, most cases report the presentation of IBD months to years before TA develops.\textsuperscript{4-11} It is uncommon to see patients with both diseases at presentation. One study includes a review of literature listing 144 cases of large vessel vasculitis, which includes TA and giant cell arteritis. Regarding IBD, 70 had Crohn’s disease and 74 had ulcerative colitis. The majority of patients (69\%) had IBD prior to their vasculitis, and 14\% had concurrent presentation.\textsuperscript{12} There are very few case reports of simultaneous presentation of pediatric TA and Crohn’s disease\textsuperscript{13-16} and no prior reports of concurrent renal artery stenosis.

Interestingly, 21\% to 36\% of patients with IBD have inflammatory processes outside of the GI system.\textsuperscript{17} The most common cardiovascular symptom is pericarditis. Others include myocarditis, thromboemboli, atrioventricular block, endocarditis, and TA.\textsuperscript{18} In a similar manner, TA could have extravascular manifestations. Those include arthritis, sacroiliitis, oral ulcers, and others.\textsuperscript{19} This overlap presents a diagnostic challenge of distinguishing consequences from coincidences. Most patients in the literature review also have systemic symptoms of inflammation, fevers, weight loss, and anorexia, similar to our patient. Another case report describes the case of a 25-year-old female who presented with GI symptoms consistent with Crohn’s disease. She later developed increasing signs of systemic inflammation, blood pressure and pulse discrepancies, and fevers. She had sclerosis of her sacroiliac joints and uveitis. Tuberculosis testing was negative. Colonoscopy revealed Crohn’s-typical ulcers. Angiography showed a plaque in the external carotid artery and thickening of the vertebral arteries, leading to a diagnosis of TA. This patient was started on steroids and methotrexate and improved.\textsuperscript{20} Her multiorgan involvement is a notable example of the overlap between the 2 diseases. Another case

Figure 2. (A) Positron emission tomography computed tomography scan demonstrating increased fluorodeoxyglucose uptake in the aortic wall, most notable just above the diaphragmatic hiatus (arrow) consistent with inflammatory vasculitis. (B) Reformatted cardiac magnetic resonance posterior oblique image demonstrating absence of head and neck vessel involvement and redemonstrating abdominal aortic narrowing (arrow).
discussed autoimmune thyroid disease in a woman who was 49 years old but had already been treated for both TA and Crohn’s disease since age 16 years.19 Our patient had renal disease and hypertension in addition to her GI and vascular symptoms—no literature could be found with a similar presentation.

One example of a simultaneous presentation is a report of a 26-year-old female who presented with systemic symptoms similar to our patient and also vomiting, abdominal pain, and watery diarrhea. She had blood pressure discrepancies on her initial examination. Her colonoscopy was consistent with Crohn’s disease, but due to her blood pressure differential, a magnetic resonance angiography revealed narrowing of the aorta and celiac artery. This led to the diagnosis of TA. The patient improved with steroids and adalimumab.21 A South African patient has also been reported to present with Crohn’s disease and TA simultaneously at 26 years of age.22 Overall, less than 8 case reports of concurrent presentation have been found in the literature.

Most literature currently involves female patients. This is expected given the higher associations of either disease with female gender. However, there were a couple of case reports of male patients. One involved the case of a 13-year-old boy who was diagnosed with TA a year after Crohn’s disease, with a new murmur and a carotid bruit on physical examination that triggered further imaging.23 Another was a case of sarcoidosis in a man who already had aortitis since age 12 years and then developed Crohn’s disease at the age of 23 years. He was treated for both diseases and responded well, but later presented with bilateral hilar adenopathy related to sarcoidosis. This unusual case report discussed the commonality of sarcoidosis to Crohn’s disease and was the first to report all 3 of the above-mentioned conditions in 1 patient. Another outlier in this article is the order of presentation. Several studies suggest HLA-B*52:01 and IL12B are with both diseases could also explain their increased concurrence.18,25 The phenotypic shared pathway could be a granulomatous intramural arteritis for both diseases.15 The idea of a common genetic component suggests a possibility of similar treatments being effective. Currently, there are multiple overlaps between IBD and vasculitis treatment: steroids, disease-modifying antirheumatic drugs, and biologic medications that target inflammatory cytokines, including tumor necrosis factor inhibitors.21 Patients with significant vascular disease sometimes need surgical or interventional radiology procedures. Our patient had angioplasty of the high-grade, short segment left renal artery stenosis. There are case reports of other vascular interventions—a repair of a saccular aneurysm of the descending aorta in a 17-year-old girl, a hemodynamically significant aortic valve insufficiency requiring replacement, and an axillary artery bypass due to stenosis.3,10,23 Most patients, however, improve with medical management and intervention is not required.

Conclusion
For patients with known IBD or those whose clinical picture includes atypical symptoms, it is important to perform thorough examinations at each clinic visit or hospitalization, including obtaining bilateral blood pressures and pulses and noting any new murmurs or bruits. Simultaneous presentation of IBD and a large vessel vasculitis are uncommon and can involve multiple other organs, resulting in unique patient presentations. Severity of disease also seems to be variable. Our patient presented with GI symptoms and renovascular hypertension. Other patients have presented with more typical presentations of decreased or absent pulses as well as presence of bruits. Currently, there are suggestions of common anti-inflammatory or anti-cytokine treatments, but research is needed to determine effectiveness of a single treatment regimen for patients who have both diseases.

Author Contributions
Dr Inna Polyakova drafted the initial manuscript, and reviewed and revised the manuscript.
Drs Kelly Rouster-Stevens, Roshan George, and Dinesh Govind Patel critically reviewed the manuscript for important intellectual content and revised it.
Drs Glen Iannucci and Anne Gill critically reviewed the manuscript for important intellectual content, revised it, and prepared figures.
All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.
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