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Takayasu Arteritis in a Child

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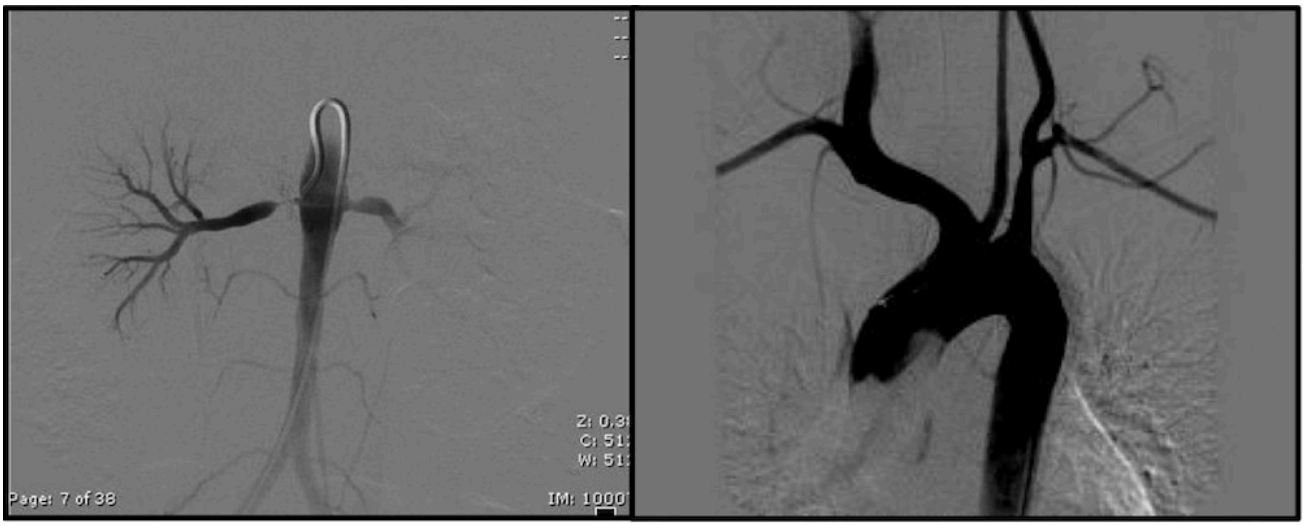
A 7-year-old girl with recently diagnosed hypertension, ulcerative colitis, and a history of Henoch-Schönlein purpura presented to the emergency department with headache, chest pain, abdominal pain, and an elevated blood pressure of 173/121 mm Hg in her right upper extremity.

Physical examination. Physical examination findings were significant for weak left carotid, brachial, and radial pulses and bilateral carotid bruits. Renal ultrasonography with Doppler was performed, the results of which were concerning for renal artery stenosis, and electrocardiography findings suggested ventricular hypertrophy.

Multiple antihypertensive agents were successively started, resulting in a slow reduction but not normalization of her blood pressure. An echocardiogram showed normal biventricular function, mild aortic root dilation, aortic insufficiency, and a diffusely dilated left coronary artery. The erythrocyte sedimentation rate (ESR) and the C-reactive protein (CRP) level were found to be elevated to 24 mm/h and 27 mg/L, respectively, raising concern for a diffuse inflammatory process.

Additional angiography of her brain, chest, and abdominal vasculature was performed, revealing significant narrowing of the left common, external, and internal carotids, and of the

revealing significant narrowing of the left common, external, and internal carotids, and of the bilateral subclavian, hepatic, splenic, and renal arteries; celiac axis narrowing; asymmetric kidney size; and diffuse thickening of the aorta from the heart through the abdomen, with a bright wall signal (**Figure**).



Diagnosis. These imaging findings, together with evidence of systemic inflammation, confirmed a diagnosis of Takayasu arteritis (TA) type V, the most extensive form. The patient was started on a methylprednisolone pulse followed by maintenance prednisone, with normalization of her ESR and CRP prior to discharge. Nevertheless, she continued to have episodic hypertension associated with headaches and abdominal pain. She subsequently underwent right renal arterioplasty as an outpatient, which led to marked symptomatic improvement and reduction of her antihypertensive regimen. She was successfully started on the disease-modifying agents methotrexate and infliximab, and her corticosteroids were tapered.

Discussion. TA is a rare yet well-described large-vessel vasculitis with a predilection for the aorta and its primary branches. The diagnosis is made clinically according to the EULAR/PRINTO/PRES criteria, which include stenosis or aneurysm of the aorta or its primary large vessel branches on angiography, not due to fibromuscular dysplasia or arteriosclerosis, as well as at least 1 of the following: hypertension, bruits, blood pressure discrepancies between limbs, pulse deficits or claudication, or elevated ESR or CRP.[1,2] The acute inflammation of TA causes nonspecific constitutional symptoms, which can lead to delay in diagnosis, and sequelae include aortic aneurysms and dissection, stricture formation, hypertension, stroke, and end-organ ischemic damage.[3-5] The etiology of TA is unknown. However, there have been case reports of TA in patients with comorbid inflammatory bowel disease,[6] such as our patient.

Treatment. Treatment options focus on preventing disease progression. There is no consensus on regimen, but high-dose pulse corticosteroid therapy is favored for induction, and long-term therapy includes immunosuppressants and biologics such as cyclophosphamide, mycophenolate mofetil, methotrexate, infliximab or tocilizumab, as well as revascularization

surgery.[4,7] Disease recurrence is common, and mortality rates can range from 16% to 40%.[7-9] Therefore, prompt identification and treatment is imperative to prevent further morbidity and mortality.

References:

1. Ozen S, Pistorio A, Iusan SM, et al, Paediatric Rheumatology International Trials Organisation (PRINTO). EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: final classification criteria. *Ann Rheum Dis.* 2010;69(5):798-806.

2. Szugye HS, Zeft AS, Spalding SJ. Takayasu arteritis in the pediatric population: a contemporary United States-based single center cohort. *Pediatr Rheumatol Online J.* 2014;12:21.

3. Cakar N, Yalcinkaya F, Duzova A, et al. Takayasu arteritis in children. *J Rheumatol*. 2008;35(5):913-919.

4. Mathew AJ, Goel R, Kumar S, Danda D. Childhood-onset Takayasu arteritis: an update. *Int J Rheum Dis*. 2016;19(2):116-126.

5. Duarte MM, Geraldes R, Sousa R, Alarcão J, Costa J. Stroke and transient ischemic attack in Takayasu's arteritis: a systematic review and meta-analysis. *J Stroke Cerebrovasc Dis*. 2016;25(4):781-791.

6. Kusunoki R, Ishihara S, Sato M, et al. Rare case of Takayasu's arteritis associated with Crohn's disease. *Intern Med*. 2011;50(15):1581-1585.

7. Ozen S, Duzova A, Bakkaloglu A, et al. Takayasu arteritis in children: preliminary experience with cyclophosphamide induction and corticosteroids followed by methotrexate. *J Pediatr*. 2007;150(1):72-76.

8. Eleftheriou D, Varnier G, Dolezalova P, McMahon A-M, Al-Obaidi M, Brogan PA. Takayasu arteritis in childhood: retrospective experience from a tertiary referral centre in the United Kingdom. *Arthritis Res Ther*. 2015;17:36.

9. Brunner J, Feldman BM, Tyrrell PN, et al. Takayasu arteritis in children and adolescents. *Rheumatology (Oxford)*. 2010;49(10):1806-1814.