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Pediatric systemic lupus erythematosus presenting with coronary arteritis: A case series and review of the literature

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ABSTRACT

Objective: Pediatric-onset systemic lupus erythematosus (pSLE) is typically more aggressive in presentation than adult-onset lupus. Presenting manifestations of lupus in children and adults involve similar organ systems, with renal and neuropsychiatric involvement more common in pSLE. Cardiac manifestations are similar in the 2 groups, with pericarditis accounting for the majority of cardiac lupus at presentation. There are no reports to our knowledge of coronary arteritis as a presenting feature of pSLE.
Methods: This is a retrospective case series describing 4 pediatric lupus patients who presented with prominent coronary artery dilatation and a review of the literature regarding coronary artery involvement in lupus.
Results: Coronary arteritis appears to be a more common feature of pSLE than previously thought. Based on our experience, coronary artery changes tend to resolve once the SLE is treated.
Conclusions: Early recognition of this disease manifestation may guide therapy and result in improved long-term cardiovascular outcomes.

Introduction

Pediatric-onset systemic lupus erythematosus (pSLE) is more aggressive than adult-onset SLE with increased severity of organ involvement. This is most often secondary to the higher frequency of renal and neuropsychiatric involvement in pSLE [1–3]. Despite these differences, pediatric and adult disease share common cardiac manifestations, primarily pericarditis, which is present in 20–30% of lupus patients at disease onset [4,5]. Other cardiac complications in pSLE include cardiomegaly, valvulitis, and conduction abnormalities [5]. Electrocardiograph changes without clinical heart disease, mitral and tricuspid valve insufficiencies, as well as left ventricular systolic and diastolic dysfunctions have also been described [5].

While coronary vessel involvement is commonly seen in patients with other systemic rheumatologic conditions, such as Takayasu arteritis, ANCA associated vasculitis, microscopic polyangiitis, and Churg–Strauss syndrome [6,7], only a few case reports have previously documented coronary arteritis in adult-onset SLE as an unusual finding [8–10]. When coronary arteritis is identified in adult-onset SLE, it is seen with vasculitis in other organ systems as well [5]. In pSLE, coronary arteritis has been reported as a rare event based on an autopsy study [11], and only 1 cohort study has indicated coronary artery dilatation in pSLE at any time point during the course of disease [12]. It would be prudent to know whether pediatric patients have coronary involvement at disease onset to assess their overall cardiovascular risk as well as to determine optimal management of their SLE.

In this article we describe 4 patients who presented with coronary artery ectasia at the onset of their diagnosis of SLE. These patients had no cardiovascular history prior to presentation. By describing these cases and reviewing the rare cases in the adult literature we seek to better identify, treat, and manage this manifestation of SLE in children.

Methods

Cases were selected from pediatric patients diagnosed with SLE and presenting with coronary artery dilatation to the Children's Hospital Los Angeles between the years 2013 and 2014. Charts were reviewed by physicians directly participating in the care of these patients. Patients underwent complete echocardiogram (ECHO) including measurements of the internal diameter of the left main coronary artery (LMCA), proximal left anterior descending coronary artery (LAD), and proximal right coronary artery

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(RCA). *Z*-scores for the coronary measurements were calculated based on pediatric normative values for body surface area built into the Xcelera R3.3 ECHO management software (Philips Healthcare, Andover, MA). The vessel was graded as dilated if the *z*-score size was greater than +2 which is >2 standard deviations above the mean. Ectasia is defined as a coronary artery being larger than normal (dilated) without a segmental aneurysm [13]. Literature review regarding other cases of SLE and coronary artery involvement was performed using a computerized Medline search of the years from 1965 to 2014. Search terms included "lupus," "pediatric," "cardiac," "coronary," and "vasculitis." In addition, a manual search of publications and cross-references was performed to obtain articles published on the subject.

Case reports

Case 1

A 9-year old Hispanic girl presented to the ED with a 4-day history of left upper quadrant abdominal pain without nausea, vomiting, or diarrhea. Her past medical history was significant for absence seizures controlled on topiramate and ethosuximide. Review of systems was notable for 2 months of arthralgias, low-grade intermittent fevers, thigh pain, and weakness of her lower extremities. She was admitted with a heart rate (HR) of 100 beats/min, respiratory rate (RR) of 24 breaths/min, temperature (*T*) of 36.1°C, and blood pressure (BP) of 114/78 mmHg. PE was notable for bilateral conjunctival erythema, a 3 mm aphthous ulcer under her tongue, tenderness to palpation of left upper and lower quadrants of the abdomen, swelling and warmth of both ankles, wrists, right knee, and several small joints of both hands. Cardiovascular exam revealed sinus tachycardia and normal heart sounds without murmurs, rubs, or gallops.

Laboratory evaluation showed mild normocytic anemia with hemoglobin (Hgb) of 11.3 g/dL (normal range: 11.5–15 g/dL), leukopenia with white blood cell count (WBC) of 3.10 k/μL (normal range: 4.5–13.5 k/μL), normal platelet count (331 k/μL, normal range: 140–400 k/μL), elevated erythrocyte sedimentation rate (ESR) of 91 mm/h (normal range: 0–15 mm/h), elevated C-reactive protein (CRP) of 5 mg/dL (normal <0.9), elevated serum lactate dehydrogenase (LDH) of 893 U/L (normal range: 140–271 U/L), positive anti-nuclear antibodies (ANA) with a homogenous pattern (1:2560), positive direct Coombs and highly elevated double-stranded DNA antibodies (anti-dsDNA >200). Additionally, positive histone antibodies were noted at 9 days post-discharge.

Her elevated inflammatory markers, fevers, and extremity swelling without frank synovitis raised suspicion for a systemic vasculitis, and an echocardiogram (ECHO) was performed to evaluate for cardiac involvement. ECHO revealed mild dilatation of the LMCA, measuring 3.8 mm (*z*-score, +2.1), and diffuse ectasia of the LAD, measuring 4 mm (*z*-score, +5.2), without discrete

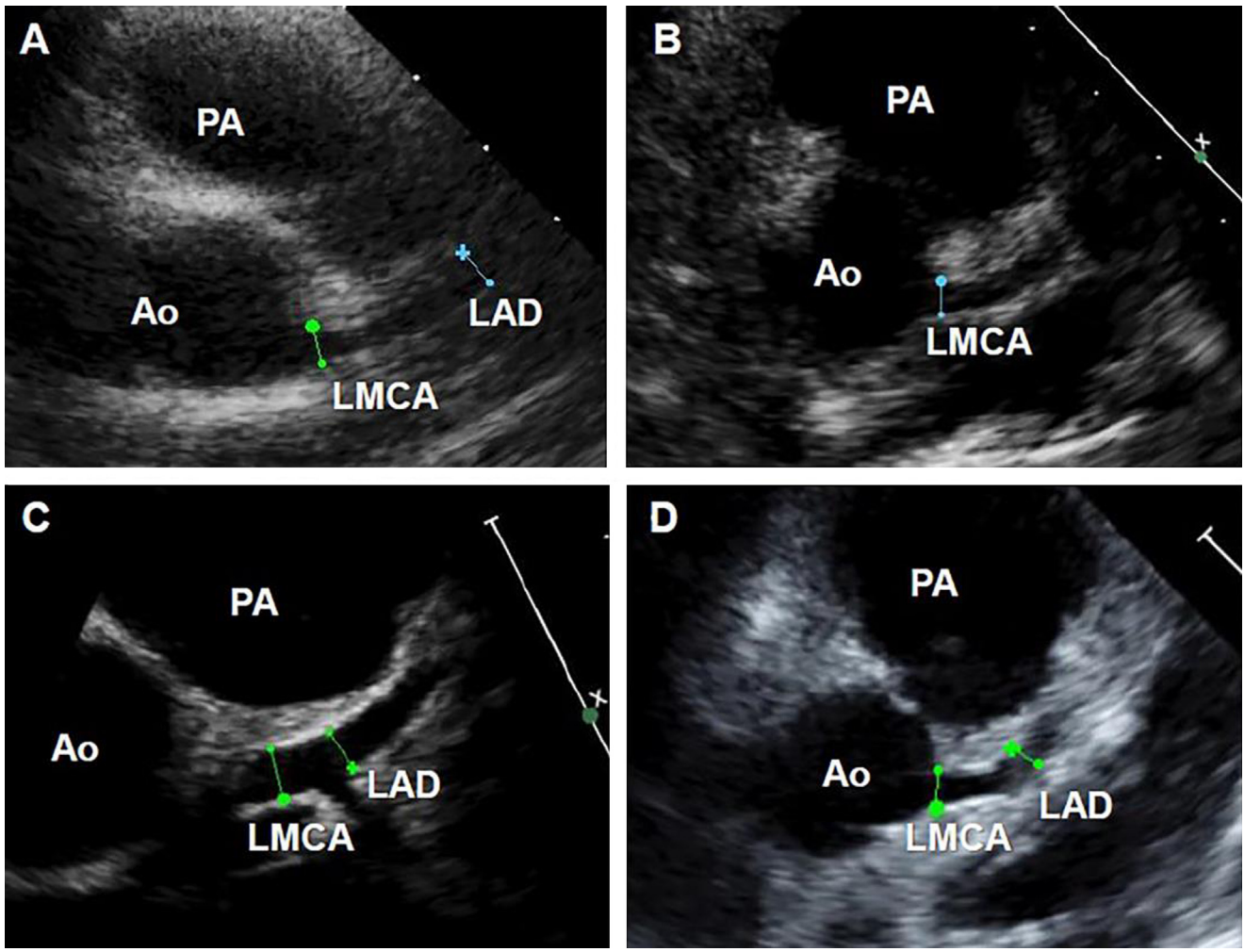

Fig. Echocardiogram stills from Case 1 (A), 2 (B), 3 (C), and 4 (D) in the parasternal short axis. (A), (C), and (D) dilated left main coronary artery (LMCA) and left anterior descending coronary artery (LAD). (B) Dilated left main coronary artery (LMCA).