Coronary Involvement in Cardiac Neonatal Lupus

A male infant, born to a mother who was receiving hydroxychloroquine because of systemic lupus erythematosus, tested positive for autoantibodies anti-SSA/Ro. His laboratory tests revealed thrombocytopenia and elevated liver enzymes; antinuclear antibodies were positive (1:160) and strongly positive for anti-SSA/Ro and anti-histone antibodies. These findings were consistent with neonatal lupus erythematosus. Echocardiography showed striking and uniform coronary artery dilatations, with right coronary artery (RCA) cross-sectional diameter being 2 mm (z score 3.5), left main coronary artery (LMCA) 2 mm (z score 2.8), and left anterior descending artery (LAD) 2 mm (z score 4.0) (**Figure**, A and B; **Video 1** and **Video 2** [available at www.jpeds.com]). Biventricular systolic function was preserved and electrocardiography found no evidence of myocardial ischemia or congenital heart block. However, an increase in cardiac markers was evident. Intravenous immunoglobulin (IVIg) has demonstrated a potential survival benefit as a treatment for autoantibody-mediated cardiomyopathy or endocardial fibroelastosis in cardiac neonatal lupus.[1-3] We therefore decided to administer IVIg infusion (2 g/kg) and started systemic corticosteroids (prednisone 2 mg/kg/d). Electrocardiograms conducted daily for follow-up remained normal, with no changes indicative of ischemia. Blood tests documented a rise in platelet count and a gradual fall of biochemical markers of myocardial necrosis. The trunk diameters of the coronary arteries were ultrasonically measured for follow-up every 3 days. We documented a gradual positive remodeling of coronary arteries, starting from RCA (**Figure**, C). The LMCA followed (**Figure**, D). Finally, after a second IVIg infusion and a parallel steroid tapering, LAD also improved. By the time the patient was discharged (25 days of age), there were no abnormal levels of platelets and cardiac and liver enzymes and coronary anomalies had completely resolved. RCA maximum trunk diameter measured 1.2 mm (z score 1.38), LMCA 1.3 mm (z score 0.43) and LAD 1.4 mm (z score 1.8) (**Figure**, E and F).

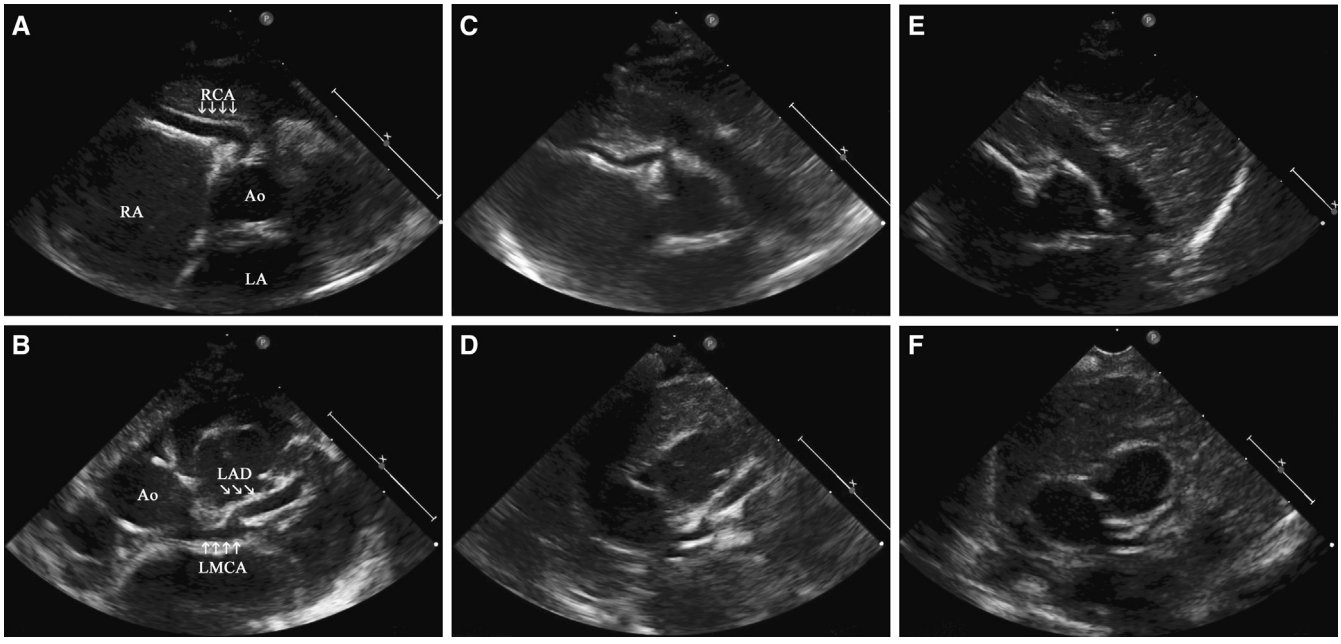

Figure. Parasternal short-axis echocardiogram view on admission showing uniform dilation of **A,** RCA and **B,** LMCA and LAD. Same view after first IVIg course and on systemic steroids, demonstrating initial positive remodeling of coronary arteries, more evident in **C,** RCA and **D,** LMCA. Parasternal short-axis echocardiogram view on discharge documenting complete resolution of coronary artery dilation, with **E,** normal RCA and **F,** LMCA and LAD cross-sectional diameters. *Ao*, aorta; *LA*, left atrium; *RA*, right atrium.

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Reported cardiac involvement in neonatal lupus, secondary to passive transplacental transfer of maternal autoantibodies to the SSA/Ro ribonucleoprotein complex, appears to be limited to fetal atrioventricular block and myocardial disease.[4] We described coronary involvement in our patient. This coronary involvement responded successfully to IVIg infusion and corticosteroid treatment. Thus, evaluation of cardiovascular complications of neonatal lupus should include echocardiographic visualization of coronary arteries, to establish early diagnosis and start prompt treatment ■.

Data Statement

Data sharing statement available at www.jpeds.com.

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References

1. Nussinovitch U, Udi N, Shoenfeld Y, Yehuda S. Intravenous immunoglobulin—indications and mechanisms in cardiovascular diseases. Autoimmun Rev 2008;7:445-52.
2. Trucco SM, Jaeggi E, Cuneo B, Moon-Grady AJ, Silverman E, Silverman N, et al. Use of intravenous gamma globulin and corticosteroids in the treatment of maternal autoantibody-mediated cardiomyopathy. J Am Coll Cardiol 2011;57:715-23.
3. Martínez-Sánchez N, Robles-Marhuenda Á, Álvarez-Doforno R, Viejo A, Antolín-Alvarado E, Deirós-Bronte L, et al. The effect of a triple therapy on maternal anti-Ro/SS-A levels associated to fetal cardiac manifestations. Autoimmun Rev 2015;14:423-8.
4. Limaye MA, Buyon JP, Cuneo BF, Mehta-Lee SS. A review of fetal and neonatal consequences of maternal systemic lupus erythematosus. Prenat Diagn, in press.

Gastrointestinal Basidiobolomycosis: A Mimic of Lymphoma

A previously healthy, well-nourished 3-year-old boy presented with fever, abdominal pain, constipation, and abdominal distension of 15 days' duration. Examination showed pallor, no peripheral lymphadenopathy, and a firm mass in the periumbilical and left lumbar areas. Laboratory investigation revealed anemia (hemoglobin 7.8 g/dL) and eosinophilia (absolute eosinophil count 3300). A computed tomography scan revealed a heterogeneously enhancing conglomerate necrotic mass encasing the jejunum, terminal ileum, ascending colon, and transverse colon (**Figure**, A). Colonoscopy identified an ulceroproliferative growth in the ascending colon and cecum with luminal narrowing (**Figure**, B). An ultrasonography-guided percutaneous biopsy specimen obtained from the mass and endoscopic mucosal biopsy specimens showed granulation tissue and eosinophilia.

The patient developed acute intestinal obstruction after 7 days of hospital stay that mandated laparotomy. The mass was unresectable on laparotomy because it involved the distal half of the small bowel and 80% of the large bowel. Thus, a venting ileostomy was performed. Surgical biopsy specimens showed dense eosinophils, giant cells, and nonseptate fungal elements displaying a Splendore–Hoeppli phenomenon (**Figure**, C). A fungal culture grew *Basidiobolus ranarum* after 4 days of incubation. An immunodeficiency workup was negative. The child had history of pica, which could have contributed to infection of the gastrointestinal tract by the fungus. He received intravenous voriconazole (4 mg/kg/day) for 6 months, followed by oral voriconazole for 12 months. Fever resolved in 2 days, and eosinophilia resolved in 2 months. The mass resolved at 15 months, after which closure of ileostomy was performed. At a 2-year follow-up, the child was asymptomatic, with normal abdominal imaging.

B ranarum is a saprophytic fungus belonging to the phylum Entomophthoromycota that usually causes chronic subcutaneous infections.[1] Gastrointestinal basidiobolomycosis is rare and may occur in individuals who are immunocompromised.[2] Presentation may mimic other diseases, such as inflammatory bowel disease and malignancies. Eosinophilia is seen in ∼65% of cases. Characteristic histopathological findings include necrotizing granulomatous inflammation, tissue eosinophilia, fungal hyphae, and evidence of a Splendore–Hoeppli phenomenon (fungal hyphae surrounded by radiating, intensely eosinophilic granular material). *B ranarum* is uniformly resistant to amphotericin B, and the drugs of choice are itraconazole and voriconazole. Prolonged therapy with antifungals is recommended for eradication.[3] ■

The authors declare no conflicts of interest.

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