

Lupus in Children Often Goes Undiagnosed

BY SUSAN LONDON

SEATTLE — The clinical features of lupus in children may be subtle and easily overlooked, Dr. David Sherry said.

Vasculitis, the pathologic hallmark of lupus, can produce a challenging clinical picture with a wide differential diagnosis, noted Dr. Sherry, who is a pediatric rheumatologist at the Children's Hospital of Philadelphia.

"It's such a black hole, a lot of clinicians just don't want to think about it." But clinicians should think about it under certain circumstances, he said.

One is when a child has constitutional symptoms that persist. "When you have a kid who is sick and they are not getting better—they still have a fever, they are still losing weight, they have an elevated sedimentation rate, and the 'virus' still isn't going away—you need to

think maybe they've got a vasculitic condition," he said at a meeting sponsored by the American Academy of Pediatrics.

Multiorgan disease can also be a tip-off of vasculitis. "Why should a kid be peeing blood and coughing up blood?" he said. "That's two different organs."

Seeing an unusual patient for the symptom, such as a teenager with a heart attack, also should raise a suspicion of vasculitis.

Finally, there is the vasculitic rash, which can have a variety of appearances.

In describing the malar rash of lupus, textbooks often show photos of a vivid, contiguous red rash in the classic butterfly distribution on the cheeks and nose, according to Dr. Sherry. But what is actually seen clinically may instead mimic rosacea, wind chapping, sunburn, or even acne. In addition, in black children, the rash may be subtle and especially hard to identify.

Key features that can help identify a malar rash of lupus include its distribution, typically with crossing over the bridge of the nose and spreading onto the cheeks, and a well-defined border between the affected skin and normal skin. Children with malar rashes usually, but not always, have other symptoms or clinical findings too.

Additional clues to the presence of lupus can often be found on parts of the body that are easily overlooked on examination, according to Dr. Sherry. For example, children may have a vasculitic rash on their hands or feet, or a painless ulcer on their hard palate. "You need to look up to see the hard palate," he pointed out. "If you look at the back of the throat, you will miss this."

The discoid rash of lupus is less common and causes crusts or scabs. "If you lift

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At the conclusion of this conference, participants will be able to:

- Identify the therapeutic options in the management of rheumatic diseases
- Explain the connection between rheumatic diseases and CV risk
- Recognize the aspects of care, treatment, and overall outcomes that are important to pediatric patients
- Describe the long-term safety and efficacy of systemic and biologic agents in the treatment of psoriasis and psoriatic arthritis
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- Recognize and describe the clinical manifestations and complications of scleroderma
- Develop a strategy for a diagnostic workup for fibromyalgia
- Discuss the challenges in managing the RA patient with IBD
- Explain the clinical manifestations and risk factors associated with gout

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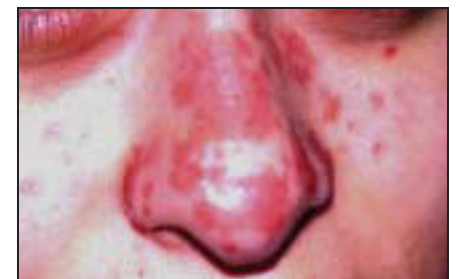
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This patient's malar rash is largely confined to the nose and isn't typical.



Children with lupus may have a vasculitic rash on their hands.



A painless oral ulcer of the hard palate can be easily overlooked.

PHOTOS COURTESY DR. DAVID D. SHERRY

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up these crusts or scabs, you see what's called carpet tacking—little pinpoints of bleeding underneath," he said. "Discoid lupus especially likes the helix of the ear, so pay attention to the helix."

Children also may have so-called lupus hairs, which are fragile and break easily. "You pull on their hair, you get three, four, or five hairs, even if they just brushed it," he explained. The breakage is accompanied by the presence of short hairs resulting from regrowth.

When lupus is first suspected in chil-

dren, Dr. Sherry recommended that physicians obtain a complete blood cell count, an erythrocyte sedimentation rate (ESR), a C-reactive protein (CRP) level, a urinalysis, a comprehensive metabolic panel, and an antinuclear antibody (ANA) titer. Lupus has the unique property of producing a high ESR and a normal CRP level—unless the child also has an infection.

An ANA panel can be deferred unless suspicion of the disease is high, he said. Related tests should be guided by symptoms, such as rheumatoid factor assessment in a child with pronounced joint

symptoms, creatine kinase assessment in a child with muscle weakness, and coagulation studies in a child with deep venous thrombosis.

Dr. Sherry noted that to be classified as having lupus, children must meet at least 4 of the 11 clinical and laboratory criteria of the American College of Rheumatology, of which a positive ANA titer is merely one.

In fact, he cautioned, 12%-20% of normal children have a positive ANA titer. "Please do not send all those normal kids with positive ANAs to your rheumatologists saying that they have lupus," he

said. "You can tell them 'You have a positive ANA.' And if you are really worried about it, we are happy to see those kids and reassure them, but ANA does not a diagnosis [of lupus] make."

If the ANA result is positive but at a titer of only 1:80 or 1:160, the child is unlikely to have lupus; if it is higher, the ANA panel should be done. "If the panel is negative, you can cool your jets and cool the mom's jets," he said. "The child doesn't have lupus."

Dr. Sherry reported that he had no conflicts of interest in association with his presentation. ■

Cholesterol Drop Precedes RA Diagnosis

COPENHAGEN — Patients who develop rheumatoid arthritis undergo an unexplained drop in their serum cholesterol level during the years immediately preceding their diagnosis, based on a study of more than 500 patients.

The finding may eventually add to a better understanding of the atherogenic process in patients with rheumatoid arthritis (RA), Dr. Elena Myasoedova said at the annual European Congress of Rheumatology. Serum levels of LDL cholesterol also fell before—as well as following—RA diagnosis, whereas serum levels of HDL cholesterol rose both before and after RA diagnosis, said Dr. Myasoedova, who did her research while she was a Fulbright scholar at the Mayo Clinic in Rochester, Minn.

Dr. Myasoedova and her associates studied residents of Olmsted County, Minn., who were enrolled in the Rochester Epidemiology Project. They identified 577 Olmsted County residents, aged 18 years or older, who were diagnosed with RA during 1988-2008.

In the 5 years before diagnosis, their serum levels of both LDL cholesterol and total cholesterol fell significantly, by an average of 24 mg/dL for LDL cholesterol and an average of 23 mg/dL for total cholesterol. During the same period, HDL cholesterol levels rose by 3 mg/dL.

In the 5 years after RA diagnosis, their total and LDL cholesterol levels underwent a smaller decline, with LDL cholesterol dropping by another 8 mg/dL. HDL cholesterol increased by an additional 5 mg/dL after the diagnosis.

To better assess the relationship of these changes to RA, the researchers ran a similar analysis on 540 control residents.

The comparison showed that people who were never diagnosed with RA also had a drop in their total and LDL cholesterol levels over the same period, Dr. Myasoedova said, but the extent of the cholesterol reduction was substantially steeper and more pronounced among those who eventually developed RA.

The study was funded in part with a grant from Roche Laboratories, Dr. Myasoedova said. She had no other disclosures for herself or her coworkers.

—Mitchel L. Zoler



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