Don't Miss CNS Vasculitis Diagnosis in Children

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CHICAGO — Pediatric vasculitis, particularly vasculitis of the central nervous system, is more common than previously recognized, according to Dr. Rayfel Schneider of the Hospital for Sick Children in Toronto.

"Each month, there seem to be more and more consults for central nervous system vaculitis," Dr. Schneider said at a meeting of the American College of Rheumatology.

Cutaneous polyarteritis nodosa (PAN) is much more common than the classic form of PAN in children, and tends to mimic juvenile rheumatoid arthritis. Dr. Schneider advises clinicians to examine the plantar surface of the feet in children suspected of having cutaneous PAN. Nodules found there are a characteristic feature. Children also present with fever, arthralgias and arthritis, splenomegaly, uveitis, and anemia.

A biopsy is needed for diagnosis, but punch biopsies do not go deep enough. The biopsy must include an artery for definitive diagnosis.

"Cutaneous PAN frequently follows a group A streptococcal infection," said Dr. Schneider. He said children may require prophylactic penicillin for recurrent streptococcal infections. Children generally respond well to prednisone, though some require other immunosuppressive therapy, particularly with organ involvement.

Kawasaki disease, which has a female predominance and a peak incidence in 1-year-old children, presents with fever for at least 5 days and at least 4 of these other clinical findings: conjunctivitis, oral mucosal inflammation, cervical lymphadenopathy, hand and foot swelling, and a rash.

Children with Kawasaki disease are also at risk of aneurysms. According to Dr. Schneider, 25% of untreated children and 5% of treated children develop coronary artery aneurysms. Children may also develop aneurysms in their axillary, brachial, iliac, and femoral arteries.

Children with acute Kawasaki disease should receive intravenous immunoglobulin (IVIG) and aspirin. Children who do not respond to this regimen may need an additional IVIG treatment, pulsed methylprednisone, or infliximab.

Because cutaneous PAN may follow a streptococcal infection, children require prophylactic penicillin for recurrent strep infections.

Henoch-Schönlein purpura (HSP) is probably the most common form of pediatric vasculitis, but when HSP presents in an atypical fashion, treating physicians should also consider Wegener's granulomatosis in the differential diagnosis. Children with Wegener's commonly present with constitutional symptoms of fever, arthralgia, and weight loss. Glomerulonephritis, upper airway disease, and lung disease also occur in 80% or more of pediatric Wegener's cases. This form of vasculitis tends to affect older female children.

In addition to vasculitis presenting with a fever and a rash, children with CNS vasculitis may present with focal neurologic deficits. Most present with acute hemiparesis, hemisensory deficits, or fine motor deficits. About half present with headaches. Findings that occur infrequently include cognitive deficits and difficulty concentrating, mood and personality changes, and seizures.

According to Dr. Schneider, serum tests, such as the sedimentation rate and complete blood counts, are frequently normal in children with CNS vasculitis. While MRI can be very sensitive in ruling out the diagnosis, cerebrospinal fluid (CSF) findings add critical information. "If you have a perfectly normal MRI and CSF, you probably don't have CNS vasculitis," he added.

Rarely, children require a brain biopsy for the diagnosis of CNS vasculitis. Children with neurologic signs and symptoms that suggest this condition, suggestive lesions on an MRI, and a normal CNS angiogram need a brain biopsy.

Testing both confirms the diagnosis of CNS vasculitis and indicates how severe it is. Children with neurocognitive dysfunction, multifocal and bilateral MRI lesions, and distal stenosis on angiogram most likely have progressive CNS vasculitis. "This is quite a devastating condition," said Dr. Schneider. ■

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