Systemic Vasculitides: Treatment Myths and Pearls

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**D ESTIN, F LA. —** Classic polyarteritis nodosa, or PAN, is curable in the majority of cases, Dr. John H. Stone said at the annual Rheumatology on the Beach. The belief that this form of vasculitis is incurable is a myth, he said. "Many patients have been told their disease is incurable," he noted. "This is a myth that needs to be debunked." Forms of the disease are tied to systemic vasculitides, he said, noting "classic PAN" means disease that does not include palpable purpura, glomerulonephritis, lung disease, or antineutrophil cytoplasmic antibody-positive disease.

In a study reported last year at the annual meeting of the American College of Rheumatology, two-thirds of PAN patients were cured, though that figure is likely high because patients other than those with classic PAN were included, noted Dr. Stone, a Boston-based rheumatologist and editor of Rheumatology UpToDate.

In his experience with classic PAN in the past 5 years, 17 of 21 patients were cured. Some were treated with steroids, and some were treated with cyclophosphamide; all were tapered completely off their drugs. The remaining four patients remained on low-dose steroids to control persistent skin disease, he said. "But we understand PAN better, we are going to recognize that we can subclassify it phenomenologically a bit more precisely," he said. Forms of the disease are tied to hepatitis B, there is the classic curable type, and there may be other subtypes.

**Episcleritis** often is an overlooked clue that a patient’s vasculitis has flared.

To debunk another myth about systemic vasculitis, Dr. Stone described a patient with Wegener’s granulomatosis who was diagnosed with rheumatoid arthritis based on the presence of arthritis and nodules on his elbows. Arthritis plus nodules does not necessarily equal RA, he said. The Churg-Strauss-type granulomas seen in this patient can be associated with Churg-Strauss syndrome, but another myth is that Churg-Strauss syndrome is the most common cause of the nodules. In fact, Wegener’s granulomatosis is the most common cause, Dr. Stone explained.

Myths also exist about treatment for vasculitis. Chemotherapy with cyclophosphamide is indicated in patients with mononeutropenia multiplex, central nervous system disease, rapidly progressing glomerulonephritis, mesenteric vasculitis, cardiac involvement, or alveolar hemorrhage. Nonhealing cutaneous ulcers or excessive steroid toxicity are also indications. Data from one study, presented only in abstract form, show that intravenous cyclophosphamide treatment once every 2 weeks is as effective as oral daily cyclophosphamide but safer. However, daily oral treatment is currently the preferred approach for induction of remission. Some studies suggest oral therapy improves the chance of sustained remission. Oral cyclophosphamide can be titered based on white cell count, another safety feature.

Patients who fail to respond to conventional daily cyclophosphamide should be considered to have an infection. Systemic vasculitis is almost always reliably controlled with conventional therapy; thus it is important to consider aspergillosis and other agents in refractory cases. Induction of neutropenia is not essential for achieving remission. The goal is to maintain white cell count above 3,500 or 4,000/μl. The current recommendation is that cyclophosphamide treatment be continued for 3-6 months after remission, followed by azathioprine. Errors with cyclophosphamide include failing to use it when needed, using it unnecessarily, and overdosing. The dose should be based on renal function and be lowered if function worsens. Elderly patients require half the dose, should have a biweekly complete blood count, receive steroids only in modulatory treatment, and receive pneumocystis pneumonia prophylaxis, Dr. Stone said.

**Vessel Size Matters in Diagnosis of Vasculitis**

**S NOWMASS , C OLO. —** The most common pitfall in diagnosing vasculitides is failure to ask about the size of the involved blood vessels, Dr. John H. Stone said at a symposium sponsored by the American College of Rheumatology. "That’s really the key to sorting out these diseases clinically," said Dr. Stone, a rheumatologist at Massachusetts General Hospital in Boston.

Large-vessel diseases can be excluded because vessels larger than 150 mcm are not present in the skin and very rarely lead to cutaneous findings. Medium-size vessels (50-150 mcm) have muscular walls, and some can be visualized. Small vessels (under 50 mcm) can’t be visualized. Vasculitides can be classified by the size of involved vessels: pure small vessel, pure medium vessel, and small/medium vessel overlap, he said.

Episcleritis may be striking, with bilateral redness of the eyes as was the case in this patient, or it can be more subtle and fleeting. But nonetheless, it is a harbinger of disease flare, Dr. Stone said.

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**Inflammatory Eye Diseases Call for Careful, Individualized Treatment to Maintain Vision**

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**C HICAGO —** Clinicians must become more aware of the prevalence and severity of inflammatory eye diseases, according to Dr. James Rosenbaum, of the Oregon Health and Science University in Portland.

"Uveitis is equal to diabetes as a cause of visual loss," said Dr. Rosenbaum at an American College of Rheumatology meeting.

Physicians may encounter several types of autoimmune uveitis in their practices. Blau syndrome, the rarest form, is an autonomic dominant disorder that causes a granulomatous uveitis and synovitis. Sometimes mistaken for sarcoidosis, it can lead to choriorretinal scarring. Another form, tubulointerstitial nephritis and uveitis (TINU), can present with systemic symptoms of fever, myalgias, and fatigue. The sedimentation rate usually is quite high. "TINU tends to be a disease of children," said Dr. Rosenbaum.

Patients respond to oral steroids. "TINU is a far more common disease than we think," he said.

In addition to uveitis, patients with systemic autoimmune disorders can present with scleritis. Dr. Rosenbaum said 40% of patients with scleritis have a systemic disease, most commonly rheumatoid arthritis (RA). Those with RA generally present with the typical features of joint pain and stiffness, and later develop scleritis. It is important to check antineutrophil cytoplasmic antibody (ANCA) levels in patients with scleritis, because the ANCA negative form is very different from the ANCA-negative form. Some medications, including bisphosphonates, can induce a local form of scleritis, but this is rare, says Dr. Rosenbaum. Withdrawing medica-