Management Is Supportive

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Additionally, inclusion body myositis has a tendency for distal and asymmetric muscle involvement, such as a foot drop. In contrast, polymyositis more commonly encompasses proximal, symmetric muscle weakness.

Pharyngeal muscle weakness is a common characteristic of both inclusion body myositis and polymyositis. In particular, however, the symptom of proximal pharyngeal spasm resulting from cricopharyngeal spasm is more often seen in cases of inclusion body myositis than in its mimic. “Patients often complain of a blocking sensation when they swallow that just doesn’t go away,” said Dr. Oddis.

Inclusion body myositis can cause marked atrophy of the quadriceps.

By comparison, the quadriceps of an unaffected individual are robust.

True Cerebral Vasculitis, Unlike Impostors, Gets Lifetime Tx

BY DIANA MAHONEY

Boston — Reversible cerebral vasocostriction syndromes resolve with simple, if any, treatment, whereas true cerebral vasculitis requires a lifetime of cytotoxic drug therapy. The clinical challenge is to distinguish the mimic from the real thing—and physicians frequently fall short of the mark, according to Leonard Calabrese, D.O.

A group of diverse conditions characterized by multifocal narrowing of the cerebral arteries, reversible cerebral vasocostriction syndromes (RCVS) are the most common and most important clinical mimic of true cerebral vasculitis, particularly primary angitis of the central nervous system (PACNS), Dr. Calabrese said at a meeting on rheumatology sponsored by Harvard Medical School.

Differentiating between the two conditions is of vital clinical importance because the respective management approaches differ substantially.

“When patients with true vasculitis of the central nervous system typically require long-term immunosuppressants and cytotoxic drugs, those with [RCVS] can often be treated with observation or calcium channel blockers alone,” said Dr. Calabrese.

This is because RCVS is self-limited, usually resolving on its own within days to weeks, whereas PACNS is a chronic, irreversible inflammatory condition.

Although both conditions share certain features, such as angiographic evidence of vasoconstriction and arteritis, variations in the clinical context provide the requisite clues for making a definitive diagnosis, noted Dr. Calabrese, head of clinical immunology at the Cleveland Clinic.

Unlike PACNS, in which there is no gender predominance, RCVS typically occurs in women aged 20-50 years. The condition often presents idiosyncratically or in a variety of clinical settings, such as after head trauma or neurovascular surgery, during pregnancy or the puerperium period, or in response to certain medications or illicit drugs, said Dr. Calabrese.

It may also occur in the setting of catecholamine-secreting tumors, he said.

“The hallmark of RCVS is the thundertap headache: extraordinarily intense, acute headache pain that crescendos with in 1 minute and has a pattern of recurring over 7-14 days,” said Dr. Calabrese. “The headache may occur with or without neurologic signs, and it may be spontaneous or precipitated by exercise, sex, coughing, or the Valsalva maneuver.”

Although headache is also the most common symptom in individuals with PACNS, “it is generally not of the acute, severe variety. Rather, headache associated with PACNS tends to be insidious and progressive, and is more often described as a dull ache.”

Because the apoplectic onset of headache in RCVS mimics subarachnoid hemorrhage, “the assumption should be that all of these patients have subarachnoid hemorrhage until proven otherwise,” Dr. Calabrese stressed. Toward this end, neuroimaging and analysis of cerebrospinal fluid are critical.

“The results of cerebrospinal fluid analysis in [RCVS] are always normal or near-normal, with no evidence of aneurysmal subarachnoid hemorrhage,” said Dr. Calabrese. In contrast, CSF examination uncovers abnormal results in more than 95% of patients with PACNS, he said, noting that the findings usually reflect aseptic meningitis, with modest pleocytosis and elevated protein levels.

Imaging studies of the brain parenchyma are normal in the majority of RCVS patients, “although MRI occasionally reveals evidence of infarction in watershed areas, parenchymal hemorrhages, small nonaneurysmal subarachnoid hemorrhages, and [posterior reversible encephalopathy syndrome],” Dr. Calabrese noted.

Dr. Calabrese mentioned that Brain MRI in true PACNS usually shows multifocal lesions in the deep white matter, and cortical infarctions in the distribution of separate vascular territories.

With respect to neurovascular imaging, RCVS cannot be differentiated from PACNS by a single cerebral angiogram. The angiographic picture that is “100% specific for vasculitis,” said Dr. Calabrese. “Angiography is most useful when the pretest probability of vasculitis is high, based on presentation and clinical symptoms.”

Although angiography occasionally reveals findings in PACNS that are characteristic of RCVS, such as nonsymmetrical vascular luminal abnormalities or excessive vascular cutoffs, the vascular abnormalities observed in PACNS patients cannot be distinguished from those seen in RCVS. The most specific finding for RCVS is evidence of substantial improvement in the characteristic vascular abnormalities—diffuse areas of multiple stenoses and dilatation involving intracranial cerebral arteries—within days or weeks of symptom onset, said Dr. Calabrese.

“In fact, the linchpin of the [RCVS] diagnosis is evidence of the reversibility of the vasocconstriction and arteritis,” said Dr. Calabrese. “The distinctive histology that you’re looking for includes endomysial inflammation, the presence of vacuoles, and intracellular amyloid deposits or twisted tubulofilaments containing hyperphosphorylated tau,” Dr. Oddis added.

Management options for inclusion body myositis are often limited to supportive efforts, for instance, myotony to relieve dysphagia caused by cricopharyngeal achalasia, said Dr. Oddis.

So far, there are no definitive treatments that have been proven effective in achieving sustained remission and improvement in a patient’s whole body strength.

However, there are some reports that suggest there may be a subgroup of patients with this disease who experience at least a partial, albeit transient response to anti-inflammatory, immunosuppressive therapy.

For this reason, said Dr. Oddis, an initial 6- to 8-week trial of prednisolone and an immunosuppressive drug such as methotrexate or azathioprine is a reasonable option for newly diagnosed patients.