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What Is the Diagnosis? Arm Weakness After T5 Paraplegia

A 38-year-old man with a 10-year history of T5 paraplegia due to a motor vehicle accident complained of acute painless swelling and limited



Image: Right-shoulder x-ray demonstrated significant destruction of the humeral head with heterotopic ossification.

range of motion of the right shoulder. He noted that his hands were numb to hot water and that phlebotomy was not painful. Physical examination revealed loss of sensation for all modalities to the T4 level, with decreased sensation of pain and temperature extending to the T2 level and also proximally in the right arm. Right-shoulder x-ray demonstrated significant destruction of the humeral head with heterotopic ossification (see image).

What is the diagnosis? Send your ideas to jwneuro@mms.org. The final diagnosis will appear in the next issue. — Sashank Prasad, MD, and Grant T. Liu, MD

Dr. Prasad is a resident in Neurology, Department of Neurology, University of Pennsylvania School of Medicine, Philadelphia.

Dr. Liu is Professor of Neurology and Ophthalmology, Division of Neuro-Ophthalmology, Department of Neurology, University of Pennsylvania School of Medicine, Philadelphia. But the relevance of these studies to clinical practice, where the goal is long-term freedom from seizures and adverse drug effects, is questionable. This non-systematic review of several newer AEDs focuses on retention rate — the percentage of patients still taking the drug after a specified period — as a reasonable measure combining efficacy and tolerability.

The author cites one study showing that at 3 years, 29% of participants remained on lamotrigine, 30% on topiramate, and <10% on gabapentin. By contrast, for levetiracetam, a study by the author demonstrated a 3-year retention rate of 37%, and a newer one that has since been published (*J Neurol Neurosurg Psychiatry* 2006; 77:101) yielded an "estimated" 3-year retention rate of 58%.

COMMENT:

The author makes a convincing case that retention rate is a reasonable measure of real-world clinical efficacy and appropriately qualifies this conclusion by advocating individualization of therapy based on comorbid conditions and pharmacokinetic properties, including

Case Diagnosis: Arm Weakness After T5 Paraplegia

In the patient with arm weakness after T5 paraplegia (see JW Neurology June 2006, p. 7), spine MRI revealed a large syrinx from C1 (image

Image 1 (above): Spine MRI revealed a large syrinx that extended from C1 (arrow). Image 2 (right): The syrinx extended to T10 (arrow).



1) to T10 (image 2), and significant kyphosis. This classic example of a Charcot neuropathic shoulder associated with post-traumatic syringomyelia likely resulted from repetitive trauma in an insensate joint as well as from dysregulated sympathetic outflow with vasomotor changes in the limb. If less severe, neurosurgical de-

> compression of the syrinx might have been beneficial. — Sashank Prasad, MD, and Grant T. Ltu, MD

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Neurology, Department of

Neurology, University of Pennsylvania School of Medicine, Philadelphia. Dr. Liu is Professor of Neurology and Ophthalmology, Division of Neuro-Ophthalmology, Department of Neurology, University of Pennsylvania School of Medicine, Philadelphia. common in industrialized nations. In this retrospective chart review from one Canadian hospital, the authors identified 12 infants admitted between 1990 and 2000 with elevated unconjugated bilirubin levels (>23.4 mg/dL) and with the pattern of brainstem and basalganglia injury suggestive of kernicterus.

Three fourths were boys, and 11 were full term. All but one had been discharged home 1 day after birth and returned at 3 to 5 days of life to the hospital with lethargy and toxic bilirubin levels. Seven had glucose-6-phosphate dehydrogenase (G6PD) deficiency, three were dehydrated, one was septic, and one had galactosemia and hemolysis. Of the children tested, three of nine had abnormal visual evoked potentials, seven of 10 had abnormal brainstem auditory evoked potentials, and five of five had abnormal EEGs. MRI suggested kernicterus in two of four, as did CT in one of five. It is not clear at what age these tests were performed. On follow-up in 10 of 12 patients (the timing and nature of which is difficult to judge), four had hypertonia, five hypotonia, three athetosis, and only two had vertical-gaze paresis.

COMMENT:

It is difficult to place the somewhat uneven data collected in this study within