Neurosurgical forum

Chiari Malformation Hydromyelia vs. Syringomyelia

TO THE EDITOR: In a recent case report (Rhoton EL, Rhoton AL Jr: Chiari malformation with syringocephaly. Case report. J Neurosurg 75:791–794, November, 1991) centered upon a case characterized clinically by hemiparesis, the authors state that this is not “the typical picture of symmetrical central cord damage commonly seen with syringomyelia.” Several points come to mind: first, the literature speaks alternatively of syringomyelia, hydromyelia, and syringohydromyelia (as a sort of compromise between the two entities). Unfortunately, the term “syringomyelia,” with its classic syndrome of central cord damage, remains emblazoned on every medical student’s mind. All of this lends further confusion to a group of lesions not clearly conceptually identical or related. In fact, in syringomyelia the cystic collection is frequently found at necropsy to meander eccentrically from the central canal, with obvious effects upon the syndromes created.

To paraphrase Sir William Osler when he said that to know syphilis in all its manifestations is to know disease, it seems to me that hydromyelia similarly presents with a remarkable panoply of neurological complexes. The perception of syringomyelia found in many old and not-so-old neurological texts seems rara avis today, and the nomenclature is obsolescent since the authors are usually imprecisely describing hydromyelia.

In the final paragraph of the Rhoton paper, the authors very briefly comment upon procedures utilized for the management of the Chiari malformation associated with hydromyelia; their own choice is the generally accepted one of decompression of the foraminal region followed by shunting of the fluid collection cavity in the cord. The statement is made that percutaneous puncture of the hydromyelic cavity (which they describe as “needling”) has been advocated as a means of treatment and discounted. In stating that leakage from the puncture site would not likely last long, the authors dismiss the possibility of valid relief.

At no time did we, in the paper to which they refer, advocate puncture of the hydromyelic cavity as primary treatment for the Chiari type syndrome; however, the procedure has been utilized with good effect for temporary (or indeterminate duration) relief of cord compression due to large fluid collections of a hydromyelic type. In speculating upon mechanisms that could account for longer-term improvement after simple cord puncture, one might consider several cogent physiological events capable of favorably influencing the hydrodynamics of cerebrospinal fluid circulation after relief of intramedullary and extramedullary compression.

Before the advent of magnetic resonance (MR) imaging, the technique of direct cord puncture also helped to delineate the true nature of the pathological changes that take place in hydromyelia over time. When visualized on multiple occasions after cord puncture and instillation of contrast agents, or by MR imaging, a picture of gradual devastation of cord structure becomes apparent, in a sort of metameric segmental pattern, as the cord loses its normal internal structural configuration. This evolution of widespread destruction of neural tissue over time manifests itself clinically in a broad spectrum of clinical presentations, mimicking syndromes as disparate as root irritation, various neuritides, and many unexpected moieties of cord involvement.

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Reference

RESPONSE: We appreciate Dr. Schlesinger’s interest in our paper as well as his previous contributions in the diagnosis and treatment of hydromyelia. The variable presentation of this syndrome with regard to motor and sensory symptomatology is well documented in the collection of 60 cases of hydromyelia presented by Dr. Schlesinger and his colleagues. The classic cervical central cord syndrome has been associated in the past with central spinal cord cavitation due to hydromyelia. This certainly represents a useful starting point in the understanding of an intramedullary spinal cord lesion. With improved imaging and treatment modalities, the frequency with which this classic syndrome is encountered is lessening. A spectrum of neurological sequelae results, depending on the direction in which the cystic cavity enlarges and destroys neural tissue.

Nineteen of the patients with hydromyelia reported by Schlesinger, et al., were treated with single or repeated percutaneous aspiration of the dilated spinal cord. In eight cases the procedures afforded lasting relief. It was concluded that percutaneous puncture and internal decompression of the hydromyelic cavities proved simple to perform and free of complications. It was considered that the neurological status of some patients could be stabilized for indeterminately long periods by simple spinal cord puncture and the removal of cerebrospinal fluid without resorting to other procedures of unpredictable efficacy. In their series, however, only 16 of the 60 patients had a Chiari type I malformation documented radiographically or at the time of operation. Aspiration of the hydromyelia associated with the Chiari malformation has been observed at the time of surgery to result in rapid refilling of the cystic cavity. It is therefore recommended that a substantial opening be made in the spinal cord, augmented with a wick to prevent future closing of the communication with the subarachnoid space. In the remaining 44 cases of Schlesinger, et al., the hydromyelic cavity was not associated with a Chiari malformation; for these patients it seems reasonable to use percutaneous aspiration as a diagnostic as well as therapeutic modality in
selected cases. This approach, however, would not be advocated in cases in which the hydromyelia is secondary to a Chiari malformation, where a more definitive treatment (foramen magnum decompression) is known to be efficacious and is aimed at the primary pathophysiologic entity as we understand it.1,2

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References

Timing of Stereotactic Procedures


We would, however, take issue with a number of their remarks and in particular with their conclusion. We find it surprising that the mean time required for stereotactic biopsy on their unit is 236 minutes (3.9 hours). In our experience of a large number of stereotactic biopsies, that figure would be much more like 60 to 90 minutes.1 This time would be even shorter if we did not have a particular interest in neuro-oncology, as a result of which we endeavor to obtain a full representation of the tumor pathology with multiple-point, single- or multiple-trajectory biopsy.

All patients undergoing biopsy, by whatever method, have an initial diagnostic CT scan, and subsequent CT for stereotactic targeting can be limited to the relevant area if CT availability is limited. In our unit, where we perform in the region of 120 stereotactic procedures a year, such scanning is part of the routine workload of the department. A neuroradiologist is not required and, on modern scanners with rapid processing and reformatting capability, the time involved is very short.

Although Di Lorenzo and colleagues are to be congratulated on their ultrasound-guided biopsy results, which are considerably better than those for freehand biopsy, their numbers are very small and the patients were selected for either stereotactic or ultrasound-guided procedures based on the size and site of the lesion. Only supratentorial lesions greater than 15 mm in diameter were chosen for ultrasound biopsy; the actual size of the larger lesions is not given and they could be very large. The patient groups are not truly comparable, therefore, and this preselection is reflected in the lesion histology; there is a preponderance of glioblastoma and astrocytoma (18 among a total of 23 cases) in the ultrasound-guided groups.

We agree that ultrasound-guided biopsy of large supratentorial lesions is to be encouraged in preference to the older freehand technique, although necessarily providing a less precise localization of particular histologic features than with CT-guided techniques. Any claims for cost saving related to the use of ultrasound rather than CT-guided stereotactic techniques must include the cost, as well as the increased risk, of repeating the procedure in negative cases.

Overall, we feel that the authors have failed to prove that the ultrasound-guided technique is in any way superior to CT-guided stereotactic biopsy other than in the debatable one of duration of the procedure.

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Reference

RESPONSE: We would like to thank Professor Hitchcock and Dr. Kenny for the interest they have shown in our article.

Expressing surprise at the mean time required for stereotactic biopsy in our center (3.9 hours), they estimate the mean time on their unit as 60 to 90 minutes. Their letter does not say whether this includes the time needed for the neuroradiological part of the procedure and their article does not clarify this point.2 We wish to specify that we timed the stereotactic procedures from the injection of the local anesthetic for headframe mounting to the patient's discharge in the recovery room. To be exact, our mean time of 3.9 hours included the following: preoperative computerized tomography (CT) scan; performing the biopsy procedure in the operating theater; postoperative CT control scan; removal of the head frame; and transferring the patient from the operating theater to the CT scan suite and back again. Patient transfers added considerably to the total time for the procedure.

In the literature, it is not easy to find exact information about the time required for performing a stereotactic biopsy. The procedure times may vary according to the head frame, the biopsy technique, and the number of targets and samples used. A recent article,1 stating stereotactic biopsy performance times calculated with the same strict criteria that we followed, gave a mean performance time of 3.7 hours, close to ours. Neurosurgical centers particularly dedicated to stereotactic surgery could probably achieve shorter times. No doubt