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MR Imaging Findings of Enteroviral Encephalomyelitis: An Outbreak in Taiwan

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In this issue of the AJNR (page 1889), Shen and colleagues describe the MR features of 20 cases of enteroviral encephalomyelitis occurring during an epidemic of hand-foot-mouth disease that swept through Taiwan in 1998. The MR findings were consistent and characteristic. The posterior medulla and pons were involved in all 15 patients with abnormal MR scans. The mesencephalon and dentate nuclei of the cerebellum were involved in two thirds of these patients. In severe cases, there was involvement of the ventral horns of the spinal cord and the deep supratentorial nuclei. The distribution of lesions on MR scans correlated well with clinical abnormalities in these patients. The majority of the children recovered clinically, and serial MR scans revealed resolution of the lesions. Two children with persistent signs of brain stem dysfunction manifested areas of brain stem and spinal cord encephalomalacia. Additional cases of severe encephalitis occurred during this epidemic. These children rapidly became comatose, suffered cardiorespiratory collapse, and died before imaging could be obtained. Based on the clinical findings, the authors postulate that these children also suffered from brain stem encephalitis with neurogenic pulmonary edema. Review of the literature reveals a strikingly similar pattern of clinical abnormalities in prior epidemics of enteroviral encephalomyelitis. The most infamous example, poliovirus, has a well-known predilection for the ventral cord and the posterior brain stem. Pathologic and imaging findings are limited but reveal the same pattern of involvement (1).

The authors conclude that the pattern of MR findings encountered in these patients is characteristic for enteroviral encephalomyelitis. In my experience, this pattern is not encountered in neoplastic, ischemic, traumatic, or metabolic disease. It is therefore reasonable to state that this pattern is more than simply characteristic; it is diagnostic of enteroviral encephalitis. [In the past, radiologists used "specific" to describe such patterns, but statisticians have usurped "specific." "Specific" now has a narrow technical meaning in scientific papers; ie, the conditional probability that a person not having a disease will be identified correctly by a diagnostic test. In order to avoid criticism and confusion, radiologists use the terms such as "diagnostic" or "characteristic." I am all for precision in scientific writing, but the loss of "specific" is a shame. It has precisely the connotation we need. I wonder if we will be forced to abandon the equally useful negation, "nonspecific." After all, if a finding cannot be specific, how can another finding be nonspecific?]

Viral encephalitis is relatively uncommon, and many patients are not imaged because they are either too sick to undergo MR imaging or too well to be thought to require it. Viral epidemics that result in encephalitis often occur in parts of the world that do not have the resources to perform neuroimaging. Therefore, imaging findings of viral encephalitis—eg, a vague paragraph in a review article or book chapter on infection, a slide pair in a refresher course lecture, some case reports in peerreviewed journals-have received little attention. Reading this literature, one comes away with a sense that these entities do not produce characteristic or consistent, let alone unique, imaging findings. There are, of course, important exceptions to this observation. Herpes simplex I and II, HIV, PML, and fetal infections have characteristic imaging features. It is no coincidence that typical imaging patterns have emerged in precisely these commonly occurring diseases that have been studied extensively. To this list of encephalitic diseases with characteristic patterns of involvement we may now add the enteroviruses.

I believe that this list will grow longer as we gain more experience with these disorders. In reality, we should follow the lead of neuropathologists and regard these diseases as individual disorders with distinctly different appearances rather than as a single disease with variable manifestations. Classifying these disorders together is based on the fact that all of these infections are characterized by diffuse and poorly circumscribed inflammation. In most other ways, these diseases differ from each other. Some infect neurons; others infect glial cells. Even within a viral group, there are different distributions owing to different modes of spread. Herpes simplex I gains access to the brain from the gasserian ganglion by growing along the meningeal branches of the trigeminal nerve and then directly invading the brain (2). Herpes zoster follows the branches of a cranial nerve (usually the fifth) back into the cavernous sinus, where it invades the walls of the carotid artery and produces a vasculitis in the distribution of its branches (3).

It seems certain that our experience with these entities will increase. Improvements in MR imaging make it easier to study seriously ill patients. Fast scanning has made it possible to evaluate the brain and spine efficiently, allowing for accurate assessment of the full extent of these diffuse diseases. Fluid-attenuated inversion-recovery and diffusion-weighted imaging have proved efficacious in the detection of subtle meningeal and superficial cortical abnormalities that can occur in meningoencephalitis. Because of these advances, MR examination of patients with relatively mild infections can reveal the full extent of CNS involvement, thus improving diagnostic accuracy. As this article illustrates, the global spread of MR technology will make it more likely that patients with epidemic encephalitis will be evaluated with MR imaging and that clinicians and scientists around the world will have access to these examinations. Globalization can have its negative effects as well. The computer virus metaphor of spread of destructive digital information is apt in the context of epidemiology. Just as these virtual viruses are spread with frightening speed over networks of connected computers around the world, biological viruses can spread by contact with the network of only slightly less mobile and connected human hosts. Diseases that were formerly isolated in remote regions of the world may invade our local populations with increasing frequency.

MR imaging promises to do more than simply document the presence of these diseases. As the authors point out, the detection of abnormalities in patients with enteroviral encephalitis may facilitate the institution of cardiothoracic support, saving the lives of children with this acute, self-limiting disease. Moreover, we have moved into an era in which neuroimaging is being used to measure outcome in drug trials. MR data has been used to evaluate the efficacy of new therapeutic agents for treatment of demyelinating disease and stroke, and it will serve the same role in the assessment of new therapeutic agents developed for viral encephalitis. These treatments will be disease-specific and will therefore require rapid and accurate diagnosis. MR imaging will surely play a major role in the process of determining the extent of involvement and the likely causative agent.

The report of a new specific pattern [there, I have said it and I am willing to bet that not one of you is confused by my usage of "specific."] is particularly satisfying because it happens so rarely now. One reason for this is the evolution of our specialty. Most of the really important patterns have already been discovered. Another reason is that this type of article has gone out of style. Articles such as this are characterized as "old fashioned" and incorrectly stigmatized as "purely descriptive." There is no quantitative data, no statistical analysis, and no direct pathologic confirmation of the hypothesis posited by Shen et al. The same can be said of the work of many scientists, including my scientific hero, Charles Darwin. Origin of the Species remains a masterpiece of scientific precision despite a complete absence of quantitative data. Some types of scientific research simply do not yield to quantitative or statistical techniques. Descriptive radiology papers have formed a critical part of our specialty because the identification of disease patterns is the basis of the practice of radiology. I find it extremely satisfying when a group of findings suddenly coalesces into a recognizable pattern-when I can look at a study and think, "I know you." I suspect that it is this pleasure that drew me, and many others, to radiology in the first place. This paper has added another recognizable pattern to the list-another disease that should not confuse us in the future.

There is another reason to pay attention to distinctive patterns. They may indicate the presence of some deeper, more general process causing the pattern. After reading this article, I believe that the marked predilection of this group of viruses for specific portions of the brain and cord must be the result of some important feature of the interaction between the virus and the brain, and that the discovery of this feature might lead to an effective treatment. As our imaging techniques improve, as we continue to obtain more detailed information on the patterns that diseases produce, we need to explore with colleagues in allied fields the deeper implications of these patterns.

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