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Spinal Cord Involvement in CNS Whipple Disease: An Ongoing Experience in the MR Imaging Era

Kremer et al (1) presented a case of Whipple disease (WD) involving the brain, optic chiasm, posterior fossa, and spinal cord. They underlined the rarity of spinal cord involvement, citing the case described by Clarke et al (2) as a unique reported case of myelopathy secondary to WD. Actually, the 62-year-old woman reported by Clarke et al had a myelopathy as a unique presentation, and MR imaging abnormalities were confined to the cord and medulla, (ie, high signal intensity on T2-weighted images, minimal contrast enhancement, and cord enlargement). Such an isolated spinal cord and medullary lesion suggested a neoplasm as one of the diagnostic possibilities, and a cord biopsy was performed; the finding of large numbers of foamy macrophages containing periodic acid-Schiff–positive bacilliform structures confirmed the diagnosis and made treatment possible. Jejunal biopsy findings were normal, but polymerase chain reaction (PCR) for Whipple's agent (*Tropheryma whippelii*) was positive.

Although myelopathy associated with WD, as a multisystem disease or confined to the CNS but involving several compartments, does not constitute a substantial diagnostic problem, an isolated spinal cord lesion in a patient without other system involvement is most likely to have this condition misdiagnosed or diagnosed late. We (3) recently described a case of severe myelopathy and an expansive spinal cord lesion highly suggestive of an intrinsic neoplasm on MR images. Biopsy was not performed; the disease had a remitting-relapsing course during a corticosteriod regimen, and only after 3 years did cerebral lesions develop. Although histologic and PCR analysis of jejunal biopsies were negative, PCR on peripheral blood finally revealed DNA of *Tropheryma whippelii*, and the clinical and imaging improvement with specific treatment was dramatic and long-lasting (at present, it persists at 22-month follow-up). To the best of our knowledge, this is the second reported case of a solely spinal presentation of CNS WD.

MR imaging shows spinal cord involvement in CNS WD as either synchronous or early and possibly isolated and appears either as a signal intensity abnormality or tumorlike lesion. A high index of suspicion should be maintained for this challenging condition, although it is rare. Cord biopsy may not be necessary, even in the case of an isolated spinal cord tumorlike lesion, because molecular biology may sometimes make the diagnosis possible in a noninvasive way.

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Recurrent Neurovascular Hypertension

We read with great interest the article by Gizewski et al (1). In this case report, the authors describe the MR findings of a patient with recurrent neurovascular compression of the left medulla oblongata.

First, we are curious to learn of the anatomic findings at the time of the first and second operations. Was (recurrent) neurovascular contact present? How was the medulla oblongata decompressed from the vertebral artery?

Second, focal edema of the brain stem at the site of compression is generally not a common finding when examining patients with essential hypertension by using MR imaging. Possibly, this group of patients with focal edema may be a distinct clinical entity and may behave differently clinically after neurovascular compression. Focal edema of the brain stem without neurovascular contact has been shown to be the likely cause of essential hypertension (2). Nevertheless, it is a well-known phenomenon that chronic pulsation, such as in cases of giant aneurysms or neurovascular contact, may result in edema and may resolve after occlusion of the artery. In the case described, the recurrence of edema may indicate recurrent neurovascular contact.

Third, the authors suggest that the clinical success might have been caused by the unintended occlusion of the left vertebral artery after the second surgical procedure. Why would this occlusion contribute to the clinical success when the artery was repositioned by fixing it to the occipital bone?

We do not agree with the suggestion that endovascular occlusion of the vertebral artery may be a primary therapy of choice. As with many patients with chronic hypertension, the vertebral artery has a high probability of being affected by atherosclerosis, and endovascular occlusion of the artery alone would not release the pressure on the medulla oblongata caused by this (atherosclerotic) artery. What if the hypertension does not improve after endovascular occlusion? Is it the result of inadequate relief of the medulla oblongata or just a lack of response to the intervention? Moreover, why put the patient at risk for delayed ischemia and the chance of being left with only one vertebral artery? In skilled hands, the operative morbidity and mortality rates associated with microvascular decompression surgery is very low (3), probably comparable with those associated with endovascular intervention.

We fully agree with the authors that MR imaging should be repeated when patients are not responsive or have recurrent hypertension. However, there is still controversy regarding whether MR imaging is a reliable tool for screening patients with essential hypertension for neurovascular contact. In this context, it is noteworthy that "positive" MR imaging findings (neurovascular contact of the vertebral artery with the left medulla oblongata) are not required for inclusion in an ongoing multicenter clinical trial of microvascular decompression for essential hypertension (4).

A substantial amount of work and research are needed to explore the true clinical effect of microvascular decompression for neurovascular hypertension. In the meantime, Gizewski et al are to be congratulated on their important and inspiring findings.

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Reply

We thank Drs. Menovsky and de Vries for their interest and comments regarding our article (1). From the surgical point of view, our reported case was an unusual case of neurovascular compression.

Because the vertebral artery was extremely ectatic, it was impossible to achieve adequate decompression of the lateral medulla oblongata by means of conventional techniques, such as interposing an implant. In both the first and second surgeries, we used a Teflon sling to transpose the vertebral artery away from the medulla oblongata. Our technique was similar to that described by Bejjani and Sekar (2). In the first procedure, the sling was fixed to the petrosal dura by using an ethilone suture. In the second procedure, which confirmed recurrent neurovascular compression, the sling was fixed transdurally to the occipital bone to achieve a more profound dorsally directed retraction. Despite these technical difficulties, we think that surgical decompression is the first choice and that occlusion or resection of a vessel is justified only in cases of failure (3).

Concerning the second-mentioned point, we agree that brain stem edema in neurovascular hypertension is an uncommon finding. We wanted to emphasize, however, that among those patients with initial brain stem edema, this aspect can be used for follow-up control and decision for reintervention in cases of recurrent edema.

Brain stem edema without vascular compression published by de Seze et al (4) was different from that in our patient. The edema in our case had close relation to the vertebral artery and did not involve the entire brain stem. We agree that edema in the brain stem may result in blood pressure dysregulation independent of the cause.

As Menovsky and de Vries note, edema as a cause of chronic pulsation, such as giant aneurysm, resolves after occlusion of the artery. We emphasize that vascular occlusion might be a possible treatment for neurovascular hypertension. This way, the pulsation is eliminated, and the vascular compression of the brain stem is diminished. In our case, vertebral artery occlusion was an unintended result but without severe neurologic complications. The primary intervention was the surgical fixation of the artery.

We also agree with Menovsky and de Vries that "positive" MR imaging findings with close contact of arteries to the brain stem are controversial. Therefore, the group of patients with initial edema, as reported in our case, represent a rare group with a potential benefit from presurgical MR imaging workup.

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Thallium-201 Single-Photon Emission CT in Recurrent Squamous Cell Carcinoma of the Head and Neck

We read with interest the report by Mukherji et al (1) concerning the use of thallium-201 single-photon emission CT to detect primary squamous cell carcinoma of the head and neck. The authors clearly show that high accuracy is obtained for thallium-201 single-photon emission CT in the differential diagnosis of recurrent tumor versus treatment effect in this tumor group, surpassing the reliability of CT in detecting this problem. We bring to the attention of your readers our work (2), which suggests another potentially important area of diagnostic benefit from thallium-201, specifically the ability to obtain prognostic information concerning the expected biological aggressivity of a childhood brain tumor. Abnormal thallium-