Symptomatic Enlarged Cervical Anterior Epidural Venous Plexus in a Patient with Marfan Syndrome

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To the Editor: We read with interest the article by Chun et al (1) in the April 2002 issue of the *AJNR*. The authors described the symptomatic enlarged cervical anterior epidural venous plexus in a patient with Marfan syndrome. They concluded that Marfan syndrome might predispose the patient to enlargement of cervical anterior epidural venous plexus secondary to a vessel wall abnormality. We would like to take this opportunity to emphasize the following point.

We draw the authors' attention to the previously published reports related to spontaneous intracranial hypotension from CSF leaks in patients with Marfan syndrome. We found five reports about this topic by conducting a MEDLINE search. However, we cannot cite all of them because of the restriction on the number of references. Schrijver et al (2) worked on 20 consecutive patients and found CSF leaks in all, four (20%) of whom exhibited minor skeletal features of Marfan syndrome. Fukutake et al (3) reported the case of a 30-year-old woman with Marfan syndrome who experienced chronic intractable headaches and spontaneous intracranial hypotension. Myelography showed multiple, large, lumbosacral arachnoid diverticula. Radioisotope cisternography revealed a halo-like accumulation in the lumbosacral region and rapid uptake of isotope in the urinary bladder, indicating CSF leakage. Epidural blood patching brought immediate relief from the headaches. Currently, anterior internal vertebral venous plexus dilation secondary intracranial hypotension is well reported in the neuroradiologic literature (4, 5).

Chun et al concluded that in their case, Marfan syndrome might have predisposed the patient to an enlargement of the anterior epidural venous plexus because of the disorder of the venous connective tissue phenomenon. However, we think that the enlargement of the anterior epidural venous plexus in their case was due to an intracranial hypotension syndrome. The patient's clinical history also supports our hypothesis.

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To the Editor: We read with interest the case report by Chun et al (1) of a patient with Marfan syndrome who presented with headache and neck pain and was found to have a large engorged cervical anterior epidural venous plexus. Chun et al suggest that Marfan disease may have predisposed the patient to an enlargement of the anterior epidural venous plexus because of a disorder of the venous connective tissue, which may be abnormal in these patients, as frequently is the arterial wall. Although we agree that venous wall abnormalities may have predisposed their patient to enlargement of the anterior epidural venous plexus, we emphasize the possible occurrence of concomitant factors due to other conditions similarly associated with Marfan disease, which might be consistent with the reportedly acute onset of symptoms. In our opinion, reduced CSF volume in the cervical region with dilatation of the cervical anterior epidural venous plexus as a compensatory phenomenon, which was suggested by the authors as an alternative hypothesis, deserves more attention as a possible causal event in the setting of such predisposition. Since the publication of Fishman and Dillon's work (2), enlargement of the anterior epidural venous plexus has been increasingly reported to occur in patients with reduced CSF volume (3), either as a feature concomitant with the more frequent MR imaging of the head findings of the so-called *intracranial hypotension syndrome* or as its most relevant or even unique feature. Growing experience with this entity has shown that its MR imaging findings may be inconstant or asynchronous. If this is the case, the difficult differential diagnosis of the enhancing epidural expanding lesion is likely to be made after invasive diagnostic procedures (4), including biopsy. Reduced CSF volume or relatively negative CSF pressure in the cervical region might be a reasonable diagnostic hypothesis for their patient with such a connective tissue disorder as Marfan disease and associated dural ectasia in the lumbar region. The findings of MR imaging of the head were reportedly unremarkable (although it is not clear whether contrast-enhanced MR images of the head were also obtained), but this may be consistent with the possible variability and lack of synchrony of MR imaging findings in cases of intracranial hypotension syndrome.

Whether the patient might have suffered from a spontaneous CSF leak or, alternatively, from pooling of the CSF in the dilated lumbar dural sac cannot be argued on the basis of the MR imaging study reported by the authors. Marfan syndrome as a connective tissue disorder is accepted to predispose to the so-called *spontaneous intracranial hypotension syndrome* caused by dural tears, which cannot be excluded in the absence of MR myelography, myelo-CT, and isotope cisternography. However, pooling of CSF in the lumbar region does also seem to be an interesting hypothesis.

In support of this, we herein briefly report our experience with a 42-year-old woman with Marfan syndrome who underwent serial imaging studies at our institution during a 15-year period. At presentation, she complained of excruciating headaches and leg pain and was found to have a large complex anterior sacral meningocele with associated intracranial findings, consistent with intracranial hypotension syndrome (ie, bilateral diffuse dural enhancement after the administration of contrast material and downward displaced midbrain structures, fourth ventricle, and cerebellar tonsils). Subsequent partial surgical treatment of the patient's extremely challenging meningocele, performed at another carefully selected institution, was successful in relieving her symptoms only for the short term, and surgical procedures to perform CSF shunting were subsequently performed as treatment of her sacral radiculopathy. Headaches continued to represent a relevant problem in her life, and on serial MR images of the head, the features of intracranial hypotension syndrome persisted over time, with the possible contribution of the otherwise useful CSF shunting. Although in this case, the intracranial hypotension syndrome is very likely to have become of mostly iatrogenic origin, its initial presentation, as associated with extensive low spinal dural ec-

tasia and conspicuous anterior sacral meningocele, shows its possible association with the neural canal disorders, which represent one of the major diagnostic criterion of Marfan syndrome (5).

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Reply

We thank Drs. Albayram, Yilmaz, and Salvolini for taking an interest in our case report of a patient with Marfan syndrome who presented with symptomatic enlargement of the anterior epidural venous plexus. Drs. Albayram and Yilmaz bring to our attention several other reports that document an association of spontaneous intracranial hypotension and Marfan syndrome. Although we do not have proof that this syndrome was present in our patient, it was among our differential diagnoses, as our article emphasized (p 623). Nonetheless, it is important to consider CSF hypovolemia as a cause of compensatory enlargement of the dural and epidural venous structures, which may appear very prominent on MR images of patients with spontaneous intracranial hypotension. We were unable to elicit a history of postural headache from our patient, and unfortunately, the patient did not undergo CSF pressure analysis; neither, however, rules this possibility out.

Dr. Salvolini also wrote to support the position of CSF hypotension as the probable cause of our patient's findings. We do not argue this point, and as we have mentioned, we did consider it to be a possible cause. He presents another case of a patient with Marfan syndrome in whom a large sacral meningocele resulted in intracranial findings of spontaneous intracranial hypotension. We thank him for the letter.

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