LETTERS TO THE EDITOR

Extrahepatic Portal-Systemic Encephalopathy without Portal Hypertension

To the Editor: We were very interested in the report of Nishimoto et al (1). Unfortunately they have obviously overlooked our manuscript (2), and their citation of previously reported cases did not appear to be correct. We reported a 76-year-old similar female case that was successfully operated and her neurological symptoms ameliorated without complications (2). Nishimoto et al stated that none of the cases were accompanied by liver cirrhosis, idiopathic portal hypertension or a history of abdominal trauma. In general, portal hypertension is defined as a state in which portal pressure is more than 200 mmH2O because the formation of various portal-systemic shunts is initiated when portal pressure exceeds that value (2, 3). At the time of publication of our manuscript, we added this standard to the same case studies as Nishimoto et al, and discovered that there have been only five cases including our patient (2, 4-7). It is unknown whether this disease is rare, and there are possibilities that other cases have been missed or just not reported. Otherwise most cases are exclusive to Japan.

Incidentally, Nishimoto et al insist on mild lymphocytic infiltration in the portal area and that considerable steatosis was the only remarkable findings in the liver biopsy of their patient. However, when we looked at Figure 3 in their article, we could note mild portal fibrosis, suspicious mild piecemeal necrosis, and intralobular lymphocytic infiltrations with suspected small focal necrosis as well. Was their case truly not accompanied by portal hypertension? The presence of portal hypertension is obscure in their case because they did not measure the portal pressure. In particular, portal pressure should be evaluated after ligation of the shunt.

Moreover, Nishimoto et al suggested that the portal-systemic shunt of their case was congenitally formed. We agree, the shunts of some cases may be congenital and related hepatic encephalopathy may occur due to increased brain sensitivity to toxic materials with aging. However, while Nishimoto et al indicated the presence of younger patients with hepatic encephalopathy from surgical portal-systemic shunt (2, 8), there have been no reports of younger cases with portal-systemic encephalopathy without portal hypertension. Therefore the congenital theory only is highly unlikely to explain all cases. From our experience I would postulate one possibility, that mild increased portal pressure theory by Watanabe et al might be in line with congenital theory. As we held a doubt about the explanation offered in previous papers for the older occurrence of encephalopathy in congenital portal-systemic venous shunt, we proposed a new hypothesis in our paper; the homeostatic control of production of ammonia might be gradually disordered with increasing age. Moreover, we speculated that some acquired changes might affect the congenital microscopic venular anastomoses, and then the anastomoses might become the functional shunt. The mild increased portal pressure theory by Watanabe et al might represent one of the acquired changes in our hypothesis, though, it is still obscure whether EPVS is congenital or not. Incidentally, we cited a report of

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Mitsunori WATANABE and Toshio FUKUSATO*

The Departments of Neurology and *Pathology, Gunma University School of Medicine, Gunma
Reprint requests should be addressed to Dr. Mitsunori Watanabe, the Department of Neurology, Gunma University School of Medicine, 3-39-22 Showa-machi, Maebashi, Gunma 371-8511

References


The authors reply:

To the Editor: We presented a 72-year-old female case of extrahepatic portosystemic venous shunt (EPVS) without portal hypertension, and reviewed previous reports as well (1). Unfortunately, we did not notice several cases as Watanabe et al pointed out. In the paper, we excluded reports of EPVS with cirrhosis, idiopathic portal hypertension or a past history of trauma in the discussion of the etiology of EPVS. Watanabe et al, moreover, ruled out cases in which portal pressure exceeded 200 mmH2O or portal pressure was not measured, to compile a strict review of EPVS (2). Indeed, it is better to measure the portal pressure so as not to include portal hypertension, but portal hypertension is not always diagnosed only by the absolute value of portal pressure. As described in our paper, we did not notice any typical, physical signs of portal hypertension such as splenomegaly, varices, ascites or anemia in our patient. Furthermore, no abnormal collateral vessels were detected by computed tomography (CT) or arteriography. It was, thus, difficult to consider that portal hypertension was present without any clinical signs, and that portal hypertension made only one portosystemic shunt open without any other collateral pathways. Therefore, we diagnosed the case clinically as EPVS without portal hypertension. Our patient and her family did not allow further examination or surgical operation at that time.

Histologically, the biopsy specimens of the liver indicated mild lymphocytic infiltration in the portal area with considerable fatty change. As Watanabe et al pointed out, mild portal fibrosis and mild piecemeal necrosis were observed in the specimens. However, these findings were obviously mild compared with those of chronic hepatitis or liver cirrhosis. We are convinced that these mild findings could not induce portal hypertension.

The etiology of EPVS, congenital or acquired, was discussed in our paper. As we held a doubt about the explanation offered in previous papers for the older occurrence of encephalopathy in congenital portosystemic venous shunt, we proposed a new hypothesis in our paper; the homeostatic control of production of ammonia might be gradually disordered with increasing age. Moreover, we speculated that some acquired changes might affect the congenital microscopic venular anastomoses, and then the anastomoses might become the functional shunt. The mild increased portal pressure theory by Watanabe et al might represent one of the acquired changes in our hypothesis, though, it is still obscure whether EPVS is congenital or not. Incidentally, we cited a report of