A Case of Marfan Syndrome Complicated with Low Cerebrospinal Fluid Pressure Syndrome

Jinya SHIOTE 1), Takako FUJITA 1), Tatsuki MIYAMOTO 1), Yukiko IHARA 1), Kazuko YOSHIMURA 1), Yukako YOSHIKANE 1), Hiroshi IDEGUCHI 1), Takahito INOUE 1), Sawa YASUMOTO 2), Shinichi HIROSE 1)

1) Department of pediatrics, Faculty of Medicine, Fukuoka University
2) Medical Education Center, Fukuoka University School of Medicine

Abstract

We report a case of the low cerebrospinal fluid pressure syndrome with Marfan syndrome (MIM 154700). The patient is a 7 year old girl, who developed headaches after falling out of bed. The headache is increased in a standing position and is relieved by lying in bed. Cranial MRI findings showed a thickening of the endocranium and an expansion of its veins; a spinal cord MRI showed an expansion of the extradural space. Because of the difficulty of collecting cerebrospinal fluid our diagnosis was low cerebral spinal fluid pressure syndrome. In addition, a diagnosis of concurrent Marfan syndrome was made. After being given rest, infusion and Angiotensin II Receptor Blocker dosage, the patient improved. It is reported that the Marfan syndrome of the adult is complicated with 17-20% of low cerebrospinal fluid pressure syndrome. However, there are few merger examples in children. A connective tissue disease may exist as an underlying disease for the low cerebrospinal fluid pressure syndrome in children. We thought that a careful medical examination was necessary.

Key words: Headache, thickening of the endocranium, Low cerebrospinal fluid pressure syndrome, Marfan syndrome