Endocrine Journal 1996, **43** (Suppl), S129–S130

A Case of Syringomyelia with Type I Arnold-Chiali Malformation (ACM): Growth Hormone (GH) Therapy and the Size of Syrinx on Serial MR Images

SATOSHI TAKAKUWA, AKIRA ASAI, AND NOBORU IGARASHI

Department of Pediatrics, Kanazawa University School of Medicine, Ishikawa 920, Japan

SEVERAL cases of hypopituitarism associated with syringomyelia and type I Arnold-Chiari malformation (ACM) have been reported [1], and in some of these cases, GH therapy might have worsened the neurological symptoms of ACM [2]. It has, however, not been fully reported that GH therapy affected the structural changes of the syrinx. In this report, we present a case of partial GH deficiency with syringomyelia and type I ACM, and show the influence of GH therapy on serial magnetic resonance (MR) images of the syrinx.

Case Study

A 10-year-old boy visited our hospital because of short stature. He had a history of a full-term normal delivery in the head position and no history of trauma. He had worn a brace for scoliosis since he was 6 years old. He complained of decreasing perspiration on the right half of his body. His height velocity was 4.3 cm/year for the previous one year. On physical examination, his height was 120.7 cm (−2.6 SD), no minor anomalies were detected, and hyperreflexia of his lower limbs was noted. His bone age was 8 years and 10 months by the Japanese TW-2 method, and the Cobb angle of scoliosis was 27 °. Endocrinological test results were as follows: peak GH levels 7.3 ng/m*l* to insulin, 6.1 ng/m*l* to arginine-HCl, and 71.4 ng/m*l* to growth hormone releasing factor; urinary GH excretion 4.8 pg/mg-Cr; somatomedin-C 322 ng/m*l*; free T_4 1.79 ng/d*l*; and other pituitary functions were within normal limits (data not shown).

At 10 years and 2 months old, GH therapy (0.5 units/kg/week) was started with sufficient informed consent because of the risk of worsening the scoliosis. Three months later, we found syringomyelia (C2-Th11) with type I ACM and normal pituitary gland on the MR image (Fig. 1A), but we continued GH therapy at his parents' request. GH therapy improved his height velocity (7.5 cm/year), and he newly began to complain of spastic gait and muscle atrophy in his right leg, and the tendon reflexes of his lower limbs were more hyperactive than before. We therefore suspended GH therapy when he was 10 years and 9 months old. Four months later, the size of the syrinx on the MR image diminished apparently (Fig. 1B). Although his height velocity decreased (3.8 cm/year) after ceasing GH therapy, and improvement in neurological symptoms and reduction of the Cobb angle (16 °) were noted.

Discussion

Recently, several cases of hypopituitarism with syringomyelia and type I ACM have been reported [1]. The pathogenesis of these abnormalities has been explained by following two mechanisms, 1) the cerebrospinal trauma by a breech delivery, 2) congenital anomaly (the midline anomaly syndrome) [1–3]. The present case with a normal delivery course could be explained by the latter mechanism.

In some of these cases, GH therapy may worsen the neurological symptoms of syringomyelia. Mori

Correspondence to: Dr. Satoshi TAKAKUWA, Department of Pediatrics, Kanazawa University School of Medicine, 13–1 Takara-machi, Kanazawa-city, Ishikawa 920, Japan
Key words: Syringomyelia, Type I Arnold-Chiari malformation, Magnetic resonance image, GH therapy

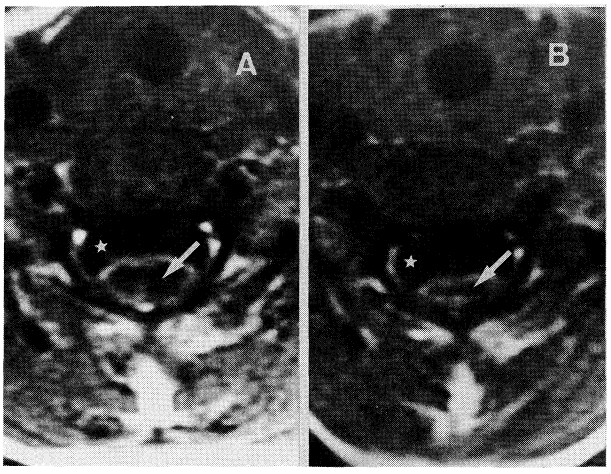

Fig. 1. Axial MR images (T1 weighted) at the C6-C7 are shown. White arrows indicate the syrinx cavity, and white stars indicate the subarachnoid space. A: This MR image 4 months after starting GH therapy shows a large syrinx cavity. Three months later, GH therapy was suspended. B: This MR image 4 months after ceasing GH therapy (7 months after A) shows a apparent reduction in the syrinx.

et al. reported that a 12-year-old boy, who had hypopituitarism with syringomyelia and type I ACM, developed such neurological complications as cerebellar ataxia, headache and anorexia 6 months after starting GH therapy [2]. Similarly, in the present case, GH replacement for 6 months may worsen the neurological signs. After ceasing GH therapy, his neurological symptoms improved with reduction of the syrinx on the MR image. It is usually assumed that the formation of syringomyelia with type I ACM is a slow progressive process. The pathogenesis in this case could be explained as follows. Accelerated growth due to GH therapy made the spinal cord stretch excessively, and the spinal cord was compressed tighter around the craniocervical junction in the presence of type I ACM (a "ball-valve" effect [3, 4]). As a result, CSF flow into the syrinx was more prominent rather than that into the subarachnoid space, and he began to complain of pyramidal tract signs. After ceasing GH therapy, impaired CSF flow into the subarachnoid space was restored and consequently the syrinx size decreased. The neurological symptoms including the Cobb angle tended to improve with reduction of the syrinx.

We can, however, not neglect the possibility of spontaneous resolution of the syrix as previously reported, especially in a post traumatic case [5]. Further follow-up studies on his clinical course are required. In the case of GH therapy for a patient with syringomyelia and ACM, close neurological examinations and serial MR studies are necessary.

References

1. Fujita K, Matsuo N, Mori O, Koda N, Okabe Y, Shirakawa N, Tamai S, Itagane Y, Hibi I (1992) The association of hypopituitarism with small pituitary, invisible pituitary stalk, type 1 Arnold-Chiari malformation, and syringomyelia in seven patients born in breech position: A further proof of birth injury theory on the pathogenesis of "idiopathic hypopituitarism". *Eur J Pediatr* 151: 266–270.
2. Mori O, Mukai E (1990) Pituitary dwarfism and syringomyelia. *Pediatr Jpn* 31: 1213–1219 (In Japanese).
3. Williams B (1980) On the pathogenesis of syringomyelia: A review. *J R Soc Med* 73: 798–806.
4. Sherman JL, Barkovich AJ, Citrin CM (1986) The MR appearance of Syringomyelia: New observations. *AJNR* 7: 985–995.
5. Jack CR, Kokmen E, Onofrio BM (1991) Spontaneous decompression of Syringomyelia: Magnetic resonance imaging findings. Case report. *J Neurosurg* 74: 283–286.