Resolution of Chiari malformation after treatment of acromegaly

Case illustration

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Chiari malformation Type I (CM-I) is defined as cerebellar tonsillar ectopia and is associated with syringomyelia in 50% of patients. We present a case of CM-I associated with cervical syringomyelia that resolved after treatment of a growth hormone (GH)-secreting pituitary adenoma.

This 39-year-old woman presented with headache and classic stigmata of GH excess. Laboratory workup revealed an elevated insulin-like growth factor–I (IGF-I) level (1120 ng/ml) and an elevated serum GH level (5.4 ng/ml) that failed to be suppressed on oral glucose testing. Magnetic resonance (MR) imaging revealed a lesion in the pituitary gland, an incidental CM-I, and cervical syringomyelia (Fig. 1). The patient underwent transsphenoidal resection of the pituitary lesion and subsequent sellar irradiation, resulting in normalization of serum IGF-I and GH levels (<1.0 ng/ml) with suppression on oral glucose testing. Histological examination confirmed a GH-secreting adenoma. At 10 months postoperatively, MR imaging revealed ascent of the cerebellar tonsils and resolution of the syrinx (Fig. 2).

There have been three reported cases of nonsyringomyelic CM-I associated with a GH adenoma in the literature.1,2,3 In only one of these was therapy directed solely at the GH excess. In that patient, clinical and neuroimaging-documented improvement of the CM-I was observed. Likewise, after GH adenoma treatment, tonsillar ectopia and syringomyelia resolved in our patient.

Acromegaly produces a well-described deformity of the acral skeleton and possibly also modifies the anatomy of the occipital bone and posterior fossa. In our case a direct relationship between the CM-I, syringomyelia, and acromegaly could not be demonstrated because no previous imaging studies had been obtained. The reduction in tonsillar herniation and resolution of the syrinx after therapy, however, may be considered indirect evidence of a causal relationship. Bone and soft-tissue deformities associated with acromegaly can partially regress after therapy. A reduction of soft-tissue thickening with alteration of cerebrospinal fluid (CSF) circulation may have occurred after GH levels normalized.1,3

Therefore, we believe that our patient’s CM-I may be attributed to GH-induced posterior fossa bone and soft-tissue hypertrophy that reduced the posterior fossa volume. This reduced volume led to tonsillar descent, abnormalities in foramen magnum CSF circulation, and syringomyelia.4 Subsequently, the correction of GH excess resulted in restoration of normal CSF pathways, tonsillar ascent, and elimination of the syringomyelia. Thus, we advocate treatment of the pituitary tumor and serial imaging to monitor for spontaneous tonsillar ascent in patients with this unusual constellation of findings.

References


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