Syringomyelia can be associated with Chiari malformation, spinal neoplasms, trauma, arachnoiditis, and other structural spinal lesions. Our patient had two potential underlying causes for a cervicothoracic syrinx: acquired Chiari I malformation and a spinal neoplasm.

This 18-year-old woman with von Hippel–Lindau (VHL) disease suffered from increasingly intense occipital headaches. Magnetic resonance (MR) imaging revealed two contrast-enhancing lesions in the right cerebellar hemisphere (Fig. 1A), herniation of the cerebellar tonsils to C-1 (Fig. 1B), and multiple contrast-enhancing lesions of the spinal cord including a ventral tumor at C-7 (Fig. 1B and C) that appeared to be associated with a cervicothoracic syrinx. Obstruction of pulsatile cerebrospinal fluid (CSF) flow at the foramen magnum was revealed on MR cine-flow studies. After suboccipital craniectomy, C-1 laminectomy, resection of the two cerebellar hemangioblastomas, and duraplasty, the patient’s headaches resolved and she has remained neurologically intact. Postoperative MR imaging and MR cine-flow studies demonstrated restored CSF flow at the foramen magnum with syrinx resolution (Fig. 1D).

The pathophysiological mechanism causing syringomyelia in this patient was related to obstruction of pulsatile CSF flow by tonsillar impaction at the foramen magnum. Treatment of the CSF flow disturbance resulted in syrinx resolution and clinical improvement and allowed for nonoperative management of spinal cord hemangioblastomas, including one next to the anterior spinal artery that proved to be asymptomatic. Because of the frequent association of syringomyelia with these hemangioblastomas of the spinal cord, the clustering of these lesions associated with VHL in the cervical segment of the spinal cord, and the high incidence of mass lesions of the cerebellum with VHL, the possibility of tonsillar impaction as the origin of cervical syringomyelia should be considered in this setting.

References