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Syrinx regression after correction of iatrogenic kyphotic deformity: illustrative case

Robert Y. North, MD, PhD, Timothy J. Yee, MD, Michael J. Strong, MD, PhD, Yamaan S. Saadeh, MD, Hugh J. L. Garton, MD, and Paul Park, MD

Department of Neurosurgery, University of Michigan, Ann Arbor, Michigan

BACKGROUND Syringomyelia has a long-established association with pediatric scoliosis, but few data exist on the relationship of syringomyelia to pediatric kyphotic deformities.

OBSERVATIONS This report reviewed a unique case of rapid and sustained regression of syringomyelia in a 13-year-old girl after surgical correction of iatrogenic kyphotic deformity.

LESSONS In cases of syringomyelia associated with acquired spinal deformity, treatment of deformity to resolve an associated subarachnoid block should be considered because it may obviate the need for direct treatment of syrinx.

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KEYWORDS postlaminectomy kyphosis; scoliosis; spinal deformity; syringomyelia; syrinx

Syringomyelia has a long-established association with spinal deformity[1–3] and is often described in relation to scoliosis. The concurrent presence of a syrinx and kyphotic spinal deformity is less often encountered.[4–10] Here we present a unique pediatric case of an enlarging syrinx at the site of prior spinal cord tumor resection. Although initially thought to be associated with the tumor, the syrinx was ultimately found to be secondary to the development of an iatrogenic, kyphotic spinal deformity. Correction of the deformity resulted in rapid and sustained regression of the syrinx. This case supports the supposition that acquired spinal deformity can contribute to syrinx formation, and in such cases, deformity correction alone may sufficiently treat both spinal deformity and the syrinx.[4,9–11]

Illustrative Case

History, Physical Examination, and Initial Imaging Findings

A 13-year-old girl with remote history of cervicothoracic anaplastic oligodendroglioma who was previously treated with multiple resections (including a C4-T4 laminectomy), adjuvant radiotherapy, and C3-L1 posterior fusion for postlaminectomy kyphosis by an outside spinal surgeon was referred to the adult spine clinic with recurrent hardware failure.

The patient was a thin (body mass index 19) but normally developed girl with a pronounced cervicothoracic kyphosis. Her neurological examination revealed mild left leg weakness, altered sensation in left arm and leg, and left lower extremity hyperreflexia. These deficits were reported as longstanding and stable. Magnetic resonance imaging (MRI) (Fig. 1) and radiographs (Fig. 2) show the patient after previous spinal fusion (Figs. 1A and 2A) performed elsewhere. At presentation to our department, MRI and radiographs revealed prominent cervicothoracic kyphosis (108°) due to hardware failure with the apex at T3-T4 and an enlarging cervicothoracic syrinx with concern for tumor recurrence (Figs. 1B and 2B; postcontrast MRI not shown). Supine imaging and flexion/extension radiographs indicated minimal correction of the kyphosis. Computed tomography revealed a posterolateral bony fusion from C3 to T3 and T4 to L1 with pseudarthrosis at T3-T4.

Operative Details

The patient was in a prone position on a flat-top surgical table and was supported with gel rolls. Her head was secured with Mayfield attachment. Multimodal neuromonitoring was used. After exposure, inspection revealed solid and confluent posterolateral fusion masses from C3 to C7 and T6 to L1. Three-dimensional navigation was used to place lateral mass screws at C3-C6 and pedicle screws at T2, T3,

ABBREVIATIONS CSF = cerebrospinal fluid; MRI = magnetic resonance imaging; POD = postoperative day.

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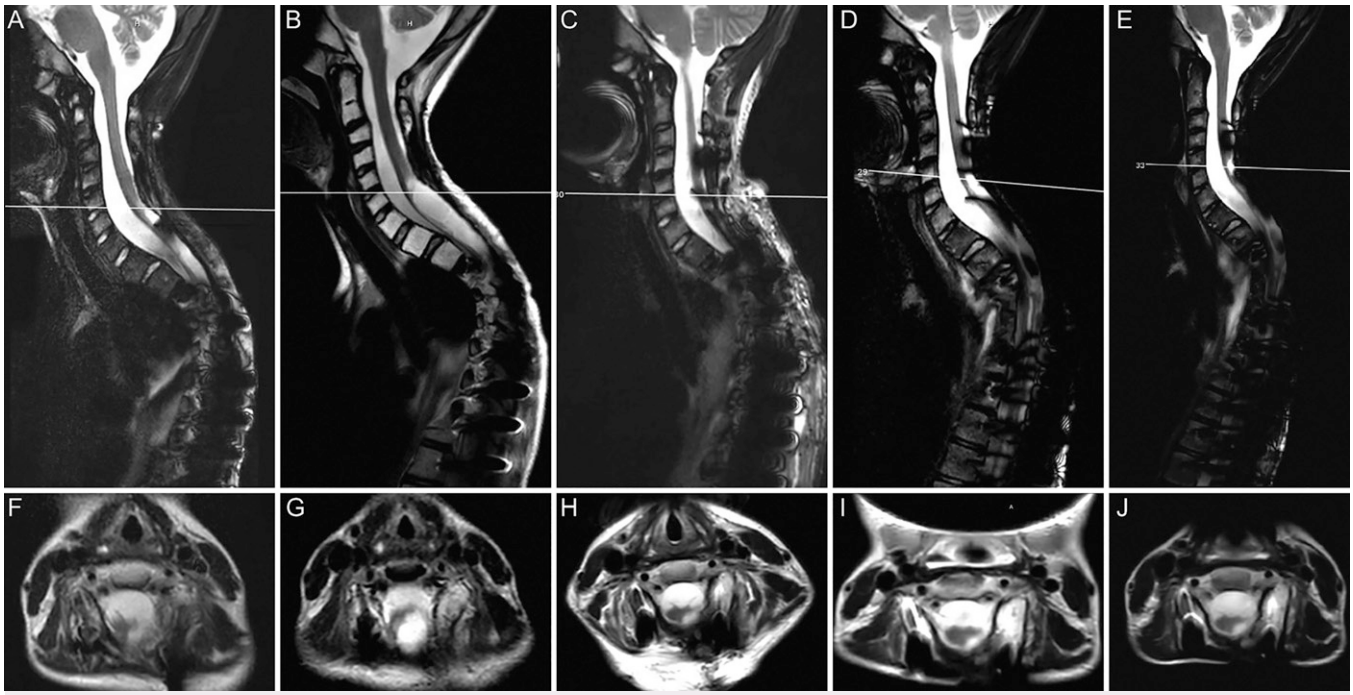

FIG. 1. Sagittal and axial T2-weighted MRI. **A and F:** After initial spinal fusion. **B and G:** After pseudarthrosis, instrumentation failure, and worsening of deformity. **C and H:** Postoperative day 1 after revision deformity correction. **D and I:** Postoperative 6-month follow-up. **E and J:** Postoperative 15-month follow-up.

and T5. Existing thoracic pedicle screws were retained. An extended T4 pedicle subtraction osteotomy, including T3-T4 discectomy and Smith-Peterson osteotomies at T2-T3 and T7-T8, was performed. After the wedge osteotomy was created, the Mayfield bed attachment was loosened and the patient's head was translated dorsally to close the osteotomy. Neuromonitoring was without changes after osteotomy closure, and fluoroscopy was used to examine sagittal and coronal alignment. Plastic surgery was performed to create bilateral paraspinous advancement flaps for closure to complete the operation.

Postoperative Course and Imaging

After surgery, the patient was neurologically unchanged from baseline. MRI and full-spine radiographs on postoperative day (POD) 1 demonstrated significant improvement in alignment (thoracic kyphosis 67°) and marked regression of cervicothoracic syrinx (Figs. 1C and 2C). The patient was discharged home on POD 3. At discharge, the patient was neurologically unchanged from baseline, ambulated without assistance, and reported minimal pain. At 3-month follow-up, the patient had improvement in

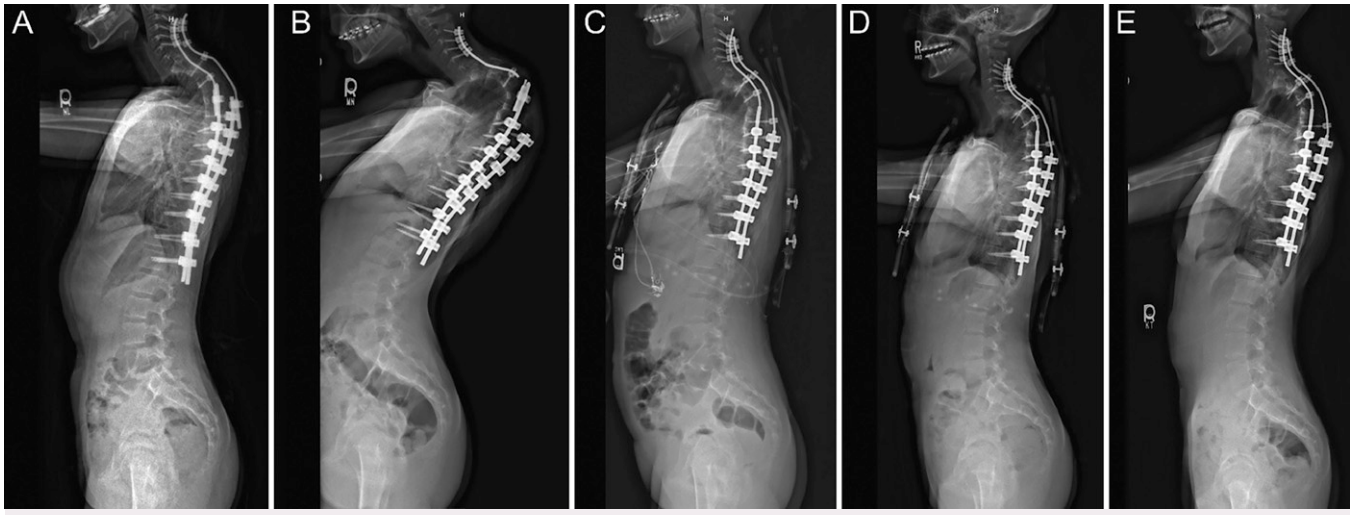

FIG. 2. Standing lateral spine radiographs. **A:** After initial spinal fusion. **B:** After pseudarthrosis, instrumentation failure, and worsening of deformity. **C:** Postoperative day 1 after revision deformity correction. **D:** Postoperative 3-month follow-up. **E:** Postoperative 15-month follow-up.

her left leg strength compared to preoperative "baseline," surgical incisions were well healed, and she reported 0/10 pain level. Full-spine radiographs at 3-month follow-up revealed sustained correction of alignment with 66° of thoracic kyphosis (Fig. 2D). Six-month follow-up MRI demonstrated stable regression of cervicothoracic syrinx (Fig. 1D). At 15-month follow-up, she reported improvement in left leg numbness, and her hypersensitivity improved. MRI demonstrated stable regression of the cervicothoracic syrinx (Fig. 1E), and upright radiographs demonstrated stable improvement of alignment without evidence of hardware failure (Fig. 2E).

Discussion

Observations

In this case, indications for surgical intervention included significant functional limitations, relative spinal instability due to partial removal of hardware, and the enlarging syrinx. The patient's skeletal maturity (Risser 5), good neurological function, and reported favorable oncological prognosis all supported surgical treatment. Preoperative imaging suggested a relatively rigid deformity, given minimal change in cervicothoracic deformity. Imaging was obtained with patient in supine position using flexion-extension radiographs or imaging in halo-gravity traction. Given the rigidity and extent of the deformity, which needed $>30°$ of sagittal correction, Schwab grade 3 or 4 osteotomy was deemed necessary.[12–16] Although theoretical estimates for sagittal correction with T4 pedicle subtraction are as high as 37.2°,[17] clinical series indicate that average correction is closer to 20° to 25°.[15,18,19] Given these estimates, additional Schwab grade 1 osteotomies were planned and proved necessary.

The etiology of syrinx formation is still unclear but is thought to be caused by disturbance in normal cerebrospinal fluid (CSF) flow dynamics.[20] For example, in Chiari I malformation, the downward herniation of the cerebellar tonsils obstructs normal CSF flow, thus resulting in an accumulation of fluid in either the central canal or spinal cord parenchyma.[20] A similar mechanism is thought to occur in other pathologies, including extramedullary lesions, infection, and trauma.[20] Tensile radial stress in the spinal cord caused by tethering is another common theory for the formation of a syrinx.[21,22] In addition to the mechanical effects of cord tethering, metabolic and ischemic changes have also been proposed as contributing factors for the formation of a syrinx in animal models.[23,24]

Regarding the syrinx in our case, our initial consideration was syrinx shunting in conjunction with deformity correction. However, when considering the temporal association of syrinx enlargement with loss of deformity correction (Figs. 1A, 1F, and 2A versus Figs. 1B, 1G, and 2B), it was thought plausible that blockage of CSF flow by the kyphotic deformity contributed to the syrinx formation and that treatment of the deformity may lead to syrinx stabilization or regression.

Lessons

In cases of syringomyelia associated with acquired spinal deformity, treatment of deformity to resolve an associated subarachnoid block should be considered because it may obviate the need for direct treatment of syrinx.

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Disclosures

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Author Contributions

Conception and design: Park, North, Saadeh, Garton. Acquisition of data: Park, North, Strong, Garton. Analysis and interpretation of data: Park, North, Strong. Drafting the article: North, Yee, Strong, Saadeh. Critically revising the article: Park, North, Yee, Saadeh. Reviewed submitted version of manuscript: Park, North, Saadeh, Garton. Approved the final version of the manuscript on behalf of all authors: Park. Study supervision: Park.

Correspondence

Paul Park: University of Michigan, Ann Arbor, MI. ppark@med.umich.edu.