Single-level ossified ligamentum flavum causing a holocord syrinx: illustrative case

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BACKGROUND Syringomyelia is a neurological disorder that is caused by abnormal cerebrospinal fluid flow or circulation. It is an incidental finding in most cases, predominantly presenting with sensory symptoms of insensitivity to pain and temperature. Spinal ossified ligamentum flavum (OLF) leading to syringomyelia is one of the rare causes. The authors report an unusual case of syringomyelia due to a thoracic OLF.

OBSERVATIONS A 54-year-old female presented with backache, difficulty walking, spasticity in the bilateral lower limbs, tingling sensation in the bilateral lower limbs, and paraparesis for 5 years. Her radiological investigations were suggestive of an OLF causing a syrinx. She underwent laminectomy, and her syrinx resolved on subsequent follow-up.

LESSONS A syrinx due to a single-level OLF is rare, and this uncommon cause should be kept in mind while formulating treatment plans.

A syrinx is a fluid-filled cavity. Anatomically, it lies within the central canal or the spinal cord parenchyma.1 Syringomyelia is a neurological disorder that is caused by abnormal cerebrospinal fluid (CSF) flow or circulation. Syringomyelia is most commonly seen in conjunction with Chiari malformation type 1.2 Syringomyelia can also be caused by extradural compressive lesions, ossified ligamentum flavum (OLF), the aftereffects of trauma, spinal cord tumor, and adhesive infectious arachnoiditis.3,4 Syringomyelia is an incidental finding in most cases, predominantly presenting with sensory symptoms of insensitivity to pain and temperature.1,5 Spinal OLF leading to syringomyelia is one of the rare causes. Cases of cervical spondylolysis leading to syringomyelia have been reported6–12. We describe an unusual case of syringomyelia due to a thoracic OLF.

Illustrative Case

History and Examination
A 54-year-old female presented with backache, difficulty walking, spasticity in the bilateral lower limbs, tingling sensation in the bilateral lower limbs, and paraparesis for 5 years. She was a homemaker and farmer by profession. She had no history of trauma or meningitis. She had not undergone any spine surgery. The power in her lower limbs progressively deteriorated from grade 5 to grade 4 over the past year. Pain sensation, temperature sensation, sense of vibration, pain, and joint position sense were diminished below the T12 level bilaterally. The patient did not have any bladder or bowel disturbances, and the reflexes were exaggerated in the bilateral lower limbs.

Imaging
Magnetic resonance imaging (MRI) of the spine (first imaging) revealed a syrinx extending from the C3 vertebral level up to the conus causing its expansion and thinning of the parenchyma. Severe impingement of the dorsal cord at the T11–12 level was noted (Fig. 1).

The patient declined surgery and was lost to follow-up because of the coronavirus disease 2019 pandemic, and she presented 6 months later with aggravated signs and symptoms. She had severe back pain, flickering movements in the bilateral lower limbs, loss of sensation below the T12 level, bladder and bowel incontinence, and severe spasticity (grade 4). MRI of the spine (second imaging) revealed an increase in the syrinx dimension with almost complete central canal dilation extending from the C3 vertebral level up to the conus and papedry thin parenchyma (Fig. 2). Computed tomography (CT) of the spine was suggestive of severe canal stenosis at the T11–12 level due to ligamentum

ABBREVIATIONS
CSF = cerebrospinal fluid; MRI = magnetic resonance imaging; OLF = ossified ligamentum flavum.

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flavum hypertrophy and ossification (Fig. 3). It was also suggestive of ossification of the interspinous ligaments.

**Treatment**

The patient underwent T11–12 decompressive laminectomy. The interspinous ligaments were calcified. An ossified and hypertrophied ligamentum flavum was identified, which was compressing the dura, and the cord was excised to achieve sufficient complete decompression of the spinal canal. No identifiable plane or dura was seen deep to the ossified and hypertrophied ligamentum flavum; hence, the cord was exposed while excising the diseased ligamentum flavum (Fig. 4). The arachnoid membrane over the posterior aspect of the cord was partially adherent to the dura.

In the postoperative period, the patient had CSF leakage (Fig. 5) from the wound, which was managed conservatively. The patient showed improvement in the postoperative period, and her symptoms improved. Paresthesia decreased in both lower extremities. Power increased to grade 3/5, and there was a significant decrease in spasticity by 1 month after surgery and to grade 4/5 by 6 months postsurgery. The histopathological examination showed ligamentum flavum ossification, significant fibrosis, and inflamed thickened arachnoid.

**Patient Informed Consent**

The necessary patient informed consent was obtained in this study.

**Discussion**

**Observations**

Primary syringomyelia due to spinal spondylolysis is rare, and only a few cases of cervical spondylolysis leading to syringomyelia have been reported.6–12 Only one case of cervical OLF resulting in syringomyelia has been reported to cause a syrinx,8 and not a
single case has been reported to cause a holocord syrinx, as was noted in our current case. We have described a rare case of dorsal ligamentum ossification leading to the formation of a holocord syrinx. Ligamentum flavum ossification can lead to myelopathy, more commonly in cases of lower thoracic spine involvement.13 OLF and hypertrophy leading to long-standing compression of the spinal cord leads to an alteration in CSF dynamics in the parenchyma of the spinal cord, thinning of the parenchyma, and enlargement of cavities due to pulsatile CSF pressure.12

Mechanical stress as an extrinsic factor is an important element in the process of the formation of OLF. Under tension stress, some fibroblastic cells can transform into chondrocytes and the expression of BMP-2, TGFβ, and SOX9 increases. Relative movement between OLF and compressed dura mater, due to flexion and extension of the spine, causes local inflammation, leading to dural adhesion. The adhesion tissue acts as a tunnel to transport osteogenic cytokines from the OLF to the compressed dura mater, which causes the dura mater directly adhered to the OLF to ossify.

Sato’s classification according to the progression of the ligament ossification is a valid tool for severity assessment and has 5 grades. Our case was Sato type D, the fused type, i.e., fusion of the bilateral ossified masses at the midline.

Resection aims at excision of the diseased segmental level and the OLF, exposing the dura for successful spinal canal decompression. No aggravated neural dysfunction or serious complications associated with this technique have been noted. Symptom duration and severity are the only factors found to be potentially predictive of surgical success.

In the present case, the patient did not have any traumatic injury or history of infection (meningitis) and had not undergone any previous spine surgery. The patient was a farmer and had had long-standing complaints. Despite all the symptoms, the patient was working until the symptoms were severely aggravated. Hence, ossified and hypertrophied ligamentum flavum at the level of spinal canal stenosis at the T11–12 level may have exposed the spinal cord at the same level to repeated minor mechanical injuries due to daily mechanical stress and exercise, thus inducing arachnoiditis, further leading to intramedullary cystic degeneration as a result of ischemia. Intramedullary cystic cavities are formed as a result of CSF pathway obstruction due to OLF.14 The on-and-off changes in the CSF pressure gradient within the spinal cord and outside the cord results in the formation of a syrinx.15

In previous reported cases of cervical spondylolysis due to OLF leading to syringomyelia, extradural decompression without any intradural surgical intervention showed significant improvement in symptoms and a decrease in the size of the syrinx.6,8–12,16,17 In this present case of dorsal ligamentum flavum ossification, we performed laminectomy and excision of the OLF (extradural decompression), but because of the long-standing spinal canal compression, the dura was adherent to the overlying ossified flavum and the arachnoid underneath the dura was adherent to the dura because of the long-standing arachnoiditis and hence was excised as part of surgery. The patient showed significant improvement in the postoperative period. Decompression of the spinal subarachnoid space along with decompression laminectomy and excision of the OLF has added beneficial effects in cases of extensive arachnoiditis, as it helps in the restoration of CSF dynamics.

Lessons

The resection of lamina and OLF is a safe and effective surgical technique in the treatment of thoracic OLF. Decompression of the
spinal subarachnoid space along with decompression laminectomy and excision of the OLFP has added benefit. We believe that durotomy is often part of the decompression surgery.

References


Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Punia, Chugh. Acquisition of data: Lachake, Gotecha. Analysis and interpretation of data: Lachake, Chugh. Drafting the article: Punia, Chugh, Gotecha. Critically revising the article: Punia, Chugh, Gotecha. Reviewed submitted version of manuscript: Gotecha. Approved the final version of the manuscript on behalf of all authors: Lachake. Administrative/technical/material support: Lachake. Study supervision: Chugh.

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