Coexisting ossification of the posterior longitudinal ligament, intramedullary hemangioblastoma, and syringomyelia of the cervical spine: illustrative case

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BACKGROUND Ossification of the posterior longitudinal ligament (OPLL) is a rare but potentially devastating cause of severe spinal cord compression and degenerative cervical myelopathy. Because OPLL is rarely accompanied by prominent syringomyelia, when both are observed, other causes of syringomyelia should be considered. Simultaneous presentation of OPLL and hemangioblastoma of the cervical spine is a rare encounter and has never been reported in the English-language literature.

OBSERVATIONS The authors present a case of a 64-year-old man with muscle weakness of the right upper limb and worsening dysesthesia of the right thumb and index finger. Noncontrast magnetic resonance imaging (MRI) of the cervical spine from another institution revealed OPLL from the C2 to C6 levels with severe spinal cord compression and prominent syringomyelia. Repeated MRI with contrast showed an intramedullary tumor, about 11 mm in diameter, at the right posterior aspect of the C4 level. The authors performed laminectomies from C1 to C6 with posterolateral fusion and removed the C4 tumor. Pathohistological examination of the tumor demonstrated hemangioblastoma.

LESSONS Careful evaluation of the preoperative imaging study is extremely important in surgical decision making. Although rare, concomitant cervical hemangioblastoma should be listed in the differential diagnosis when OPLL is accompanied with prominent syringomyelia.

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KEYWORDS case report; cervical spine; hemangioblastoma; ossification of the posterior longitudinal ligament; syringomyelia

Ossification of the posterior longitudinal ligament (OPLL) is a rare but potentially devastating cause of severe spinal cord compression and degenerative cervical myelopathy. When the spinal cord is severely compressed and the flow of cerebrospinal fluid is obstructed, syringomyelia can occur. Possible causes of syringomyelia are several, but most cases are associated with Chiari malformation, then spinal cord injury, meningitis/arachnoiditis, tethered cord syndrome, and spinal tumors. Because OPLL is rarely accompanied with prominent syringomyelia, when both are observed at the same time, whether there are other possible causes of syringomyelia should be reconsidered. Simultaneous presentation of OPLL and hemangioblastoma of the cervical spine is a scarce encounter and has never been reported in the English literature. In this case report, we present a case with coexisting OPLL, intramedullary hemangioblastoma, and prominent syringomyelia of the cervical spine.

Illustrative Case

A 64-year-old male came to our outpatient department for a second opinion about the surgical choices to treat his OPLL at the cervical spine to determine whether an anterior, posterior, or combined approach suited him better. He presented with muscle weakness of the right upper limb...
and worsening dysesthesia of the right thumb and index finger lasting for months. Unsteady gait and hyperreflexia of the four limbs were also noted. Noncontrast magnetic resonance imaging (MRI) of the cervical spine from another institute revealed obvious OPLL from the C2 to C6 levels with severe spinal cord compression and prominent syringomyelia (Fig. 1). Careful evaluation of the noncontrast MRI study, however, revealed a suspicious intramedullary tumorous lesion at the C4 level (Fig. 1C). Repeated MRI with contrast was therefore arranged, which showed, as expected, a well-enhanced intramedullary tumor, about 11 mm in diameter, at the right posterior aspect of the spinal cord (Fig. 2A and B). The differential diagnoses included ependymoma, astrocytoma, hemangioblastoma, intramedullary metastasis, and lymphoma, etc. Preoperative computed tomography of the cervical spine showed segmental OPLL at the C2–6 levels, causing severe spinal stenosis. A highly vascularized tumor was also noticed at the right posterior aspect of the C4 level.

Under the impression of C2-6 OPLL and a C4 intramedullary tumor, a posterior approach was adopted. We performed laminectomies from C1 to C6 with posterolateral fusion and removed the C4 tumor. Under the microscope, a solid reddish tumor with abundant vascular supply was encountered (Fig. 3). There was severe adhesion of the tumor to the surrounding spinal cord. Resection of the tumor was carried out with gentle dissection and bipolar coagulation. Pathohistological examination of the tumor was consistent with the diagnosis of hemangioblastoma (World Health Organization grade I): proliferation of capillaries with variable sizes, closely packed vessels, and large neoplastic stromal cells (Fig. 4). The patient had transient postoperative allodynia over his right forearm and hand. He could carry out daily activities without assistance and was discharged from the hospital uneventfully. At the 3-month follow-up, his handgrip strength and dexterity of the right hand had improved to the degree that he could use chopsticks. His gait also improved gradually.

Discussion
Observations
The incidence of OPLL in the general population varies, with the highest reported prevalence being in East Asian countries. The
preoperative embolization in fear of horrendous complications. Well enhanced on MRI, our neuroradiologist did not recommend an intramedullary tumor was suspected. Despite the tumor being about the surgical choices for OPLL. After careful evaluation of the radiologists. In our case, the patient came for a second opinion with multidisciplinary discussion among neurosurgeons and neuro-embolization was only recommended in those highly selected cases. There was also embolic agent penetrating into the tumor in one of them, which gave the tumor a hard consistency along with its arte-travasation of the embolic agent was observed in two cases, and subsequent unilateral cerebellar syndrome. In their study, minor ex-vascular spinal cord tumors: report of five cases and review of the literature. AJNR Am J Neuroradiol. 2005;26(4):936–945.
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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: Liao, C-R Li, Shen. Acquisition of data: Liao, C-R Li, Lee, H-N Li. Analysis and interpretation of data: Liao, C-R Li, Lee, Shen. Drafting the article: Liao, C-R Li. Lee. Critically revising the article: Liao, Cheng, Shen. Reviewed submitted version of manuscript: Liao, Cheng. Approved the final version of the manuscript on behalf of all authors: Liao. Study supervision: Liao.

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