Extensive ossification of the paraspinal ligaments in a patient with vitamin D-resistant rickets: Case report with literature review

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ABSTRACT

INTRODUCTION: Ectopic ossification of the spinal ligaments is not uncommon in patients with Vitamin D-resistant rickets (VDDR), but the long-term consequences of this condition have not been reported.

PRESENTATION OF CASE: The case was a 65-year-old female with VDDR who reported progressive weakness of the upper extremities, difficulty walking, neck pain, and numbness in the left arm. Imaging studies demonstrated cord compression with ectopic ossification at the rim of the occipital bone and OPLL at C1 level. Ankylosis of the whole spine below the C2 vertebrae was also noted with preserved mobility only at the craniovertebral junction.

DISCUSSION: Our report showed that ectopic ossification of the spinal ligament can result in ankylosis of the entire spine in patients with VDDR. In such patients, the segments with remaining mobility are considered to be at high risk of developing myelopathy due to increased stress at the junction.

CONCLUSION: The present case underscores the importance of performing long-term follow-up in VDDR patients presenting with ectopic ossification of the spinal ligaments. In particular, physicians should pay close attention to the possibility of myelopathy in any segments with preserved mobility.

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1. Introduction

Vitamin D-resistant rickets (VDDR) is a group of metabolic disorders characterized by renal tubular defects in phosphate transport and bone abnormalities resulting in hypophosphatemic rickets or osteomalacia. Patients with VDDR may present with deformities of the lower extremities, bone or joint pain, short stature or dental abnormalities and tend to develop calcium deposits at the attachments of tendons or ligaments as they age. It is recognized that ectopic ossifications of the spinal ligaments are not uncommon in VDDR patients, which may result in compression of the spinal cord and subsequent myelopathy. Despite this, the long-term consequences of ossified spinal ligaments in VDDR patients have not been well documented. We report a case of VDDR presenting with extensive ossification of the spinal ligaments for which we performed long-term follow-up.

2. Presentation of case

The patient was first referred to our hospital because of difficulty in walking at the age of 34, when she was diagnosed with VDDR. Later, the diagnosis was genetically confirmed as described by a different research group.

The patient had previously undergone T7–T9 laminectomy due to thoracic myelopathy at another hospital at the age of 24, after which her myelopathic symptoms subsided for 7 years. At the age of 34, she underwent a second posterior decompression surgery (T4–T9) for gait disturbance due to thoracic myelopathy after a diagnosis of OPLL and OYL, which resulted in improvement of her symptoms. Since then, the patient has been followed-up on annual basis and remained functionally stable for over 30 years.

At the age of 65, she reported weakness of the upper extremities, difficulty walking, neck pain, and numbness in the left arm. She was admitted for further investigation and treatment. On admission, she was 118 cm tall with a marked round back and bowed legs. She was able to walk only short distances supporting herself on a wall. Neurologic examination revealed decreased light touch and pinprick sensation, and motor weakness (3/5 strength) in the distal upper extremities. The grip power was 6 kg in both hands. Tendon reflexes were equivocal with indifferent Babinski sign bilaterally. Plain radiograph showed marked kyphosis of the thoracic spine (T1–T12 angle: 94°) (Fig. 1). Computed tomography demonstrated ankylosis of the whole spine below the C2 vertebra with extensive ossification of the paraspinal ligaments (Fig. 2A, B). In contrast, decreased but preserved mobility (9° on flexion and extension) was noted at the craniovertebral junction (CVJ). No overt radiographic instability was found in the atlantoaxial region, with an atlantoaxial interval of 1 mm. In addition, there was ossification at the...
Fig. 1. Plain lateral radiograph showing marked kyphosis of the thoracic spine.

Table 1
24 cases of VDRR patients with myelopathy caused by intracanal pathology.

| Authors, year      | Age(yr)/sex | Follow-up duration | Intracanal pathology (level) | Surgery (level) |
|--------------------|-------------|--------------------|------------------------------|-----------------|
| Dugger and Vandiver [7], 1966 | 60/M | N/A | bony overgrowth (T11/12) | laminectomy (T11, 12) |
| Johnson et al. [8], 1966 | 28/F | 2.5 month | bony overgrowth (T11) | laminectomy (T10-L1) |
| Yoshikawa et al. [9], 1968 | 48/F | 6 months | OPLL (C2-4) | laminectomy (C2-5) |
| Highman et al. [10], 1970 | 28/M | 15 months | thickened laminae (C2-5) | laminectomy (C2-5) |
|                      | 55/M | 6 months | thickened laminae (T6-8) | laminectomy (T6-8) |
| Bradbury et al. [11], 1987 | 50/F | N/A | OLF (T5/6, 6/7, 7/8) | laminectomy (T5-8) |
|                      | 42/M | N/A | OPLL (T5-8), OLF | laminectomy (C3-T1, T7, 8) |
| Matsui et al. [3], 1991 | 39/M | 7 years | OLF (T7-10) | laminectomy (T7-11) |
| Bussiere et al. [12], 1993 | 49/M | N/A | OLF (T8/9) | laminectomy (T8-10) |
|                      | 57/M | N/A | OLF (T5-8) | laminectomy (T3-9) |
| Yamamoto and Onofrio [13], 1994 | 42/F | 6 months | degenerative changes (T3-11) | laminectomy (T3-11) |
| Ballantyne and Findlay [14], 1996 | 32/M | 3 months | OLF (T7-11) | laminectomy (T7-11) |
|                      | 45/M | 6 months | CLF (T10/11) | laminectomy (T10, 11) |
| Dunlop and Stirling [15], 1996 | 49/F | 3 months | OLF (T5/6, 6/7, 7/8, 9/10) | laminectomy (T6-10) |
| Vera et al. [16], 1997 | 48/M | 5 years | OLF (Thoracic including T9/10) | laminectomy (T10) |
|                      | 40/F | N/A | CLF (C5/6) | laminectomy (C2-6) |
|                      | 43/F | N/A | CLF (cervical) | laminectomy (cervical) |
| Velan et al. [5], 2001 | 54/F | 2 years | OPLL (C4-6) | laminoplasty (C3-7) |
| Soehle and Casey [4], 2002 | 44/F | N/A | OPLL (C2, 3, 4), calcified disk (C6/7) | anterior surgery (C6/7) |
| Kawaguchi et al. [18], 2009 | 44/F | 5 years | OPLL (C5-T1, T12/L1), OLF (T8/9, T9/10, T10/11) | laminoplasty (C2-7, T8-11), anterior surgery (T12/L1) |
| Lee et al. [2], 2012 | 36/F | 3 years | OPLL (C1/2, C2/3-6/7) | laminectomy and fusion (C3-7) |
| Shiba et al. [19], 2015 | 32/F | 2 years | OPLL (C2-7) | laminoplasty (C2-6) |
| Present case | 65/F | 31 years | ossification at the rim of the occipital bone, OPLL (C1/2) | resection of both ossified rim of the occipital bone and posterior arch of C1 |

VDRR, Vitamin D-resistant rickets; N/A, data not available; OPLL, ossification of the posterior longitudinal ligament; OLF, ossification of ligamentum flavum; CLF, calcification of ligamentum flavum.
myelopathy. Similar pathomechanisms have been reported in patients with ankylosing spondylitis, presenting as various pathologies at the craniovertebral junction including atlantoaxial subluxation, retroodontoid pannus, basilar invagination, and bone erosion [20].

This study was reported in line with the CARE criteria [21].

Fig. 2. Sagittal reconstruction images of computed tomography demonstrating fusion of the entire spine below C2: Cervical spine (A) and thoracolumbar spine (B).

Fig. 3. T2-weighted magnetic resonance image revealing spinal cord compression at the C1 level by the ossified tissues.

4. Conclusion

The present case underscores the importance of continuing the periodic monitoring of VDRR patients presenting with ectopic ossification of the spinal ligaments over a period of decades. Physicians should pay close attention to possible myelopathy occurring at the segment with preserved mobility.

Conflict of interest

The authors declare that they have no conflict of interest.

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Ethical approval

For this type of study formal consent is not required.

Patient consent

Informed consent was obtained from all individual participants included in this study.

Author contribution

Yujiro Hirao: contributed to data collection and writing the manuscript. Hirotaka Chikuda: was the supervisor and contributed to discussion and writing the manuscript. Yasushi Oshima: contributed to discussion and data collection. Yoshitaka Matsubayashi: contributed to discussion and data collection. Sakae Tanaka: contributed to the discussion.
