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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 (VDRR), but the long-term consequences of this condition have not been reported.
PRESENTATION OF CASE: The case was a 65-year-old female with VDRR 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 vertebra 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 VDRR. 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 providing long-term follow-up in VDRR 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.

1. Introduction

Vitamin D-resistant rickets (VDRR) is a group of metabolic disorders characterized by renal tubular defects in phosphate transport and bone abnormalities resulting in hypophosphatemic rickets or osteomalacia[1]. Patients with VDRR may present with deformities of the lower extremities, bone or joint pain, short stature or dental abnormalities [1] and tend to develop calcium deposits at the attachments of tendons or ligaments as they age [1]. It is recognized that ectopic ossifications of the spinal ligaments are not uncommon in VDRR patients, which may result in compression of the spinal cord and subsequent myelopathy [2–5]. Despite this, the long-term consequences of ossified spinal ligaments in VDRR patients have not been well documented. We report a case of VDRR 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 [6].

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 atlantodental interval of 1 mm. In addition, there was ossification at the

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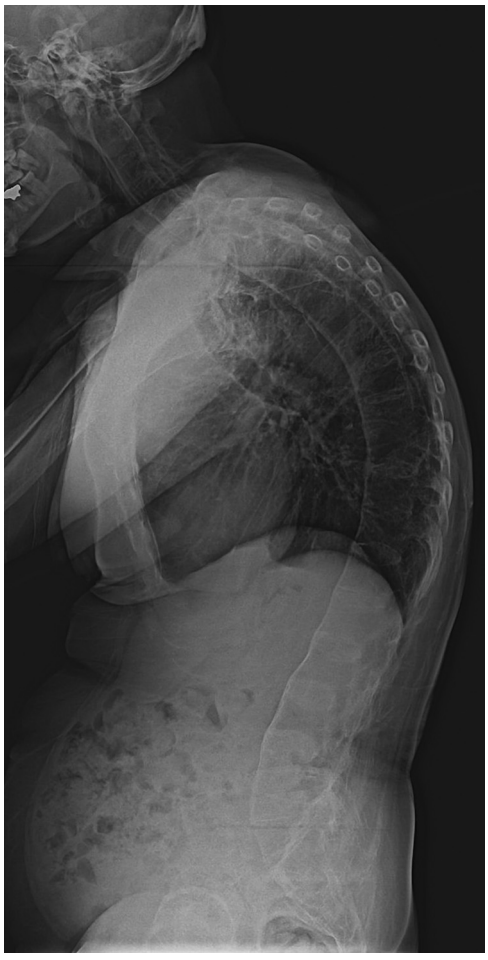

Fig. 1. Plain lateral radiograph showing marked kyphosis of the thoracic spine.

rim of the occipital bone, and OPLL at the C1 level. Magnetic resonance imaging (MRI) revealed spinal cord compression at the levels of both the occipital bone and C1 (Fig. 3). The patient underwent posterior decompression, in which the posterior arch of C1 and the ossified rim of the occipital bone were resected. Deformation of the dural sac was observed at locations corresponding to the resected portions of the rim of the occipital bone and C1 posterior arch, indicating that there had been sustained pressure on the sac. Her postoperative course was uneventful. At the 18-month follow-up visit, the patient was free of pain and numbness. Her grip power had improved to 20 kg in the right and 15 kg in the left hand. She had regained the ability to walk with the support of a cane.

3. Discussion

In this report, we described a case of VDRR associated with extensive ossification of the spinal ligaments followed up for more than 30 years. Ossification of spinal ligaments has been reported in approximately one third of VDRR patients over the age of thirty, and is thought to be linked to both genetic and environmental factors [2,5,6]. In the literature, 23 cases of VDRR patients undergoing surgical treatment for myelopathy have been reported (Table 1) [2,3,7–19]. The majority of the patients are middle-aged (mean age, 44.3 years). In most cases, surgical decompression resulted in favorable outcomes in terms of neurological recovery. However, in reports that consider the implications for spine surgery or changes in the radiological findings, most of the reported follow-up periods are 2 years or under and the longest period reported is 7 years. In this case, further problems arose 30 years after surgical decompression, a period that has not previously been covered.

In the present case, ossification of the spinal ligaments progressed to ankylosis of the entire spine below C2, mimicking ankylosing spondylitis. Given the preserved but minor mobility at the craniovertebral junction, continuing mechanical stress may have been imposed on the stenotic lesion, leading to

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 (C3–4)	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)
	55/M	N/A	OLF (T5/6, 6/7, 7/8)	laminectomy (T5–8)
Bradbury et al. [11], 1987	50/F	N/A	OLF (C3–5, T7/8)	laminectomy (C3–T1, T7, 8)
	42/M	N/A	OPLL (C3–5, T7/8), OLF	anterior surgery (C3/4, C4/5), laminectomy (C3–7)
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)
Thomas and Burnet [17], 2000	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, C4), 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