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## Case report

## Hirayama-like disease in the thoracic spine ☆

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## ABSTRACT

Hirayama disease is a cervical flexion myelopathy that typically causes upper extremity weakness in young male patients. We present two male patients (age 15 and 29) with MRI findings of thoracic ligamentous laxity similar in appearance to Hirayama disease. However, patients presented with atypical symptoms, specifically back pain and paresthesia of the upper and/or lower extremities, likely correlating to the abnormal thoracic spinal levels involved. Flexion/extension MRI sequences demonstrated the forward displacement of the dorsal dura and compression the thoracic cord with prominence of the posterior epidural space and venous plexus. Follow-up MRAs were negative for a spinal vascular malformation. Patients were managed conservatively with no surgical intervention. Clinical history, thoracic MRI, and follow-up flexion and angiographic imaging sequences may help confirm a diagnosis of Hirayama-like thoracic ligamentous laxity.

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## Introduction

Hirayama disease is a flexion myelopathy that involves the lower cervical segmental myotomes, typically C5-6 or C8-T1, resulting in upper extremity weakness in young male patients [1–3]. Cervical spinal cord injury in Hirayama disease may be secondary to abnormal dynamic compression of the cord by surrounding structures, with forward displacement of the dorsal dura compressing the cervical cord, as seen on flexion MRI sequences [1,4,5].

Characteristic cervical MRI findings of Hirayama disease include lower cervical spinal cord atrophy and T2WI hyperintense signal, loss of the cervical lordosis, loss of the dural attachment to the lamina with forward displacement on flexion MRI, and prominence of the posterior epidural space and venous plexus [1,2,6].

We present 2 patients with imaging features similar to Hirayama disease in the thoracic spine using MRI and assess imaging and clinical follow-up for these patients.

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**Fig 1 – Case 1: Follow-up flexion/extension thoracic MRI demonstrate forward migration of the posterior wall of the dura mater and accentuation of the dorsal epidural venous plexus (arrows) at the level of T2-T3 to mid T7 on (A) extension and (B) flexion sequences. (C) Follow-up MRA was negative for underlying arteriovenous fistula.**

## Case series

### Case 1

A 29-year-old male with type I diabetes, pancreatic insufficiency, scoliosis, and winged scapula initially presented with one year of thoracic pain after a back flip into a pool. Four years later at 33 years of age, the thoracic pain extended to bilateral legs and shoulders with occasional arm involvement. Pain was aggravated with back extension. He also reported lower extremity paresthesias and weakness, loss of color below the knees bilaterally, and hand numbness and coldness. On physical exam the thoracic spine was tender to palpation; otherwise, general and neurological physical exams were normal. The patient had low vitamin D levels (22.1 ng/mL), normal rheumatological lab values, and a normal vitamin B12 level (737 pg/mL).

On MRI, the thoracic spine demonstrated reversed s-shaped scoliosis. There was prominent dorsal epidural fat and venous plexus from T3-6. Two-month follow-up flexion/extension MRI demonstrated forward migration of the posterior wall of the dura mater and accentuation of the dorsal epidural venous plexus at the T2-T7 levels on flexion (Figs. 1A and B). Laxity of the ligamentum flavum, in addition

to mild degenerative disc disease, resulted in mild to moderate spinal canal stenosis at these levels. Follow-up MRA with and without contrast was negative for underlying vascular malformation (Fig. 1C).

An imaging differential of atypical Hirayama's like disease in the thoracic spine was offered given appearance on flexion/extension sequences with a negative MRA. On clinical re-evaluation, the patient demonstrated no findings of myelopathy. Surgery was consulted with no intervention recommended. Patient was treated for low vitamin D and managed conservatively with trigger point injections. The patient continued to have pain two years later at age 35.

### Case 2

A 15-year-old male initially presented with Lhermitte sign. Specifically, the patient reported 1-2 years of tingling and burning sensations in the mid to lower back extending along the back of the legs to the knees when he flexed his neck forward. General and neurological physical exams were normal. Laboratory follow-up was not obtained.

Follow-up thoracic spine MRI demonstrated a non-enhancing T2WI hyperintense focus within the spinal cord at T3 (Fig. 2A). Prominent dorsal epidural fat and venous plexus were noted at levels T1-10. There was associated mild



**Fig 2 – Case 2: Follow-up thoracic MRI obtained at 15 years of age demonstrates (A) abnormal T2WI hyperintensity and volume loss of the cord at T3 (arrowhead). (B) Extension and (C) flexion sequences demonstrate increased enlargement of the epidural venous plexus of T1-10 and anterior displacement of the spinal cord (arrows). Same day MRA was negative for underlying arteriovenous fistula (not shown). (D) Follow-up MRI and MRA at 22 years of age demonstrated persistent abnormal T2WI hyperintensity, progressive volume loss (arrowhead), and unchanged prominent dorsal epidural venous plexus (arrow).**

flattening of the posterior lateral cord at T3-4. Follow-up one-month MRA was negative for a vascular malformation. Flexion/extension sequences obtained at the time of the MRA demonstrated mild enlargement of the venous plexus and anterior displacement of the spinal cord with flexion (Figs. 2B and C). Findings were not significantly changed on follow-up MRIs at age 16. At age 17, the T2WI hyperintense intramedullary signal at T3 was less conspicuous on follow-up MRI and MRA. At age 22, MRI spine demonstrated progressive volume loss of the spinal cord at T3 and persistent abnormal T2WI hyperintense signal compared to initial imaging (Fig. 2D).

An imaging differential of atypical Hirayama's like disease in the thoracic spine was offered given appearance on flexion and extension sequences with a negative MRA. Surgery was consulted but no intervention was recommended. At age 17, the patient was re-evaluated by neurosurgery and found to be asymptomatic with a normal physical exam. On most recent follow-up at age 22, the patient remained asymptomatic.

## Discussion

We present two cases of ligamentous laxity of the spine on MRI with imaging features similar to Hirayama disease al-

though located in the thoracic spine. The exact pathophysiology of Hirayama disease is unknown but may be due to insufficient growth of the dura causing ischemia of the anterior horn cells in the lower cervical spine as the displaced shortened dura compresses the spinal cord on flexion [1,7]. While compressive myelopathy from ligamentum flavum laxity in the lower thoracic spine has been described in patients with trisomy 21, it has not been widely reported in other patients [8]. Loss of cervical dural attachment may have 70-93% sensitivity and 100% specificity for Hirayama [1,6]. In both our cases, detachment of the dura over multiple levels may have compressed the thoracic spinal cord with flexion, causing symptoms.

Hirayama disease in the cervical spine typically occurs in young male patients who present with asymmetric upper extremity muscle weakness and atrophy [1]. At initial presentation, both patients were young (age 15 and 29) and male. Presenting symptoms were variable in our two cases and included pain and/or paresthesia of the upper extremities, shoulders, lower back, and/or lower legs/feet. However, patients' symptoms were centered in the lower extremities and aggravated with certain movements. Compared to the typical presentation of upper extremity weakness in Hirayama, atypical symptoms in our cases were likely related to the lower anatomic distribution of abnormal ligamentous laxity in the thoracic spine. Possible myelopathy with abnormal T2WI

signal was seen in the second patient. This finding may be less frequent in the thoracic spine because the spinal canal is more capacious, and the spinal cord is relatively smaller at this level compared to the cervical spine.

Diagnostic imaging features of Hirayama disease in the cervical spine include detachment of the dura from the lamina, and forward displacement of the dura with flexion [1]. Both patients demonstrated similar MRI findings, but in the thoracic spine. Patients with Hirayama may also demonstrate lower cervical cord atrophy, and abnormal cervical cord T2WI hyperintensity [1], which was only seen in the second patient at the level of the T3 vertebral body. Loss of the normal cervical lordosis has also been described in patients with Hirayama disease [1]. Our first patient demonstrated abnormal thoracic alignment with reversed s-shaped scoliosis. Imaging findings of Hirayama are typically identified in the cervical spine [1], although one case of Hirayama in the literature described prominence of the epidural space and venous plexus at C5-6 and T2-8 with abnormal T2WI signal at C6 [9]. Unlike previously described cases of Hirayama disease, imaging abnormalities in our cases were centered in the upper thoracic spine with variable extension inferiorly.

Despite atypical clinical presentations and thoracic imaging findings, Hirayama-like disease was included in the differential as a diagnosis of exclusion. Differential diagnosis also included arteriovenous fistula. Imaging follow-up for Hirayama includes cervical MRI flexion and extension sequences [1], and was obtained for both patients. Flexion and extension sequences demonstrated enlargement of the venous plexus with displacement of the spinal cord and/or dorsal dura. Follow-up MRA imaging was negative for arteriovenous fistula in our two cases, upholding a diagnosis of Hirayama-like disease in the thoracic spine. Symptoms of Hirayama typically stabilize with conservative management with a neck collar [1,9], and surgery is rarely warranted [3]. Both patients were managed conservatively with no surgical intervention, and the second patient's symptoms completely resolved four years after his initial symptoms.

In summary, dorsal ligamentous laxity in the thoracic spine can be seen in a rare subset of patients presenting with atypical symptoms of myelopathy or radiculopathy attributable to the upper thoracic spine. The etiology may be similar to Hirayama disease in the cervical spine, in that ligamentous laxity may compress the upper thoracic cord with motion. As with Hirayama disease, if thoracic ligamentous laxity is suspected, flexion sequences are recommended as they may confirm detachment of the dura with increased prominence of the dorsal epidural space on flexion. Follow-up MRA or catheter angiography can be performed if concern for vascular anomaly persists [10].

## Patient consent

All data in this case report are retrospective and all patient identifiers were removed. Patient consent was not obtained.

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