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Multilevel Spinal Canal Stenosis with Ossification of the Ligamentum Flavum in An Achondroplastic Patient: A case report

***Arya Widyatama and Adam Moeljono**

Departement of Orthopaedic Surgery, Academic Hospital, Gadjah Mada University, Yogyakarta, Indonesia
*Corresponding author: aryawidy97@yahoo.com

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Abstract

Introduction: Achondroplasia is a bone disorder that causes dwarfism due to genetic disorders, particularly autosomal dominant mutations in Fibroblast Growth Factor Receptor 3 (FGFR 3). Mutation of FGFR 3 resulting in decreased growth plate size, reduced trabecular bone volume, and decreased bone elongation influenced neurologic and pain problem. Early surgical decompression is the key to gain better neurologic recovery and pain improvement among patients with achondroplasia**. Case:** It was reported a 23 years old man suffered from low back pain and bilateral weakness of both lower extremities for about 8 months followed by intermittent neurogenic claudication and motoric function loss problems two days later. Patient also came with mild scoliosis, reduced interpediclar distance of thoracal spine, and several spinal canal stenosis at L2-L5 level of lumbar spine accompanied by ossification of the ligamentum flavum. Wide decompressive laminectomy with spinous process-splitting approach and resection of the Ossification of Ligamentum Flavum (OLF) were performed at L2 to L5 level using a high-speed burr and a Kerrison**.**

Conclusion: Decompressive wide laminectomy and resection of adherent OLF followed by fusion with instrumentation were performed using the same technique as we mentioned above. No dural tear was recorded on this patient. There was an improvement on patient's neurological symptoms and function after the surgery. This method is safe and effective.

Keywords: Achondroplastic, spinal canal stenosis, ossification

1. Introduction

Achondroplasia is a bone disorder caused by genetic problem that begins in early fetal development so the growth of cartilage into the appropriate bone is inhibited. Achondroplasia is the most common form of dwarfism in humans. It occurs with a frequency of 1 in 15-25,000 and 80% of cases are sporadic. It is caused by autosomal dominant mutations in Fibroblast Growth Factor Receptor 3 (FGFR 3). FGFR 3 is expressed in chondrocytes and mature osteoblast where it functions to regulate bone growth. Increased of FGFR 3 induce suppresses proliferation and maturation of growth plate chondrocyte resulting in decreased growth plate size, reduced trabecular bone volume, and decreased bone elongation (1).

2. Case

A 23 years old man suffered from low back pain and bilateral weakness of both lower extremities for about 8 months. He fell down after slipped in front of his bathroom with

bottocks hit the first contact on the floor, April 2021. Two days later, intermittent neurogenic claudication appeared, he started to feel pain on his lower back, loss of sensation on his lower part of the legs (below knee) and medial side of his toes, also loss of motoric function on both of his legs. He has history of hydrocephalus and underwent surgery when he was a kid.

Figure 1. Preoperative image of 23-year-old man with large head, prominent forehead, short arms and legs with long trunk

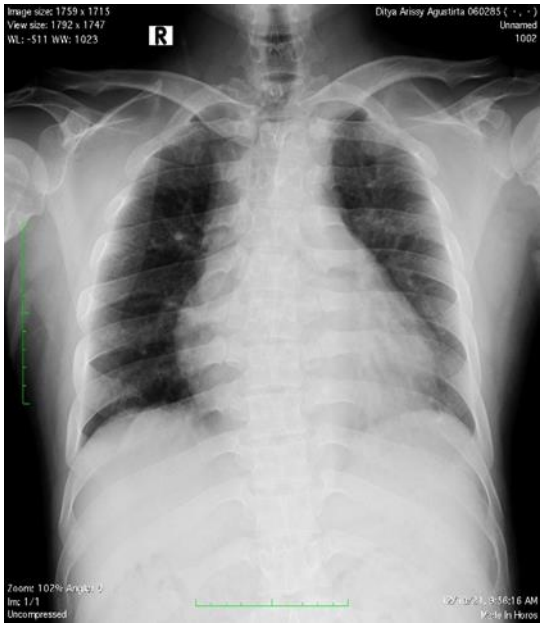

Figure 2. Anteroposterior radiographs of the thoracolumbar spine demonstrate mild thoracic scoliosis, with decreased inter-pedicle distance

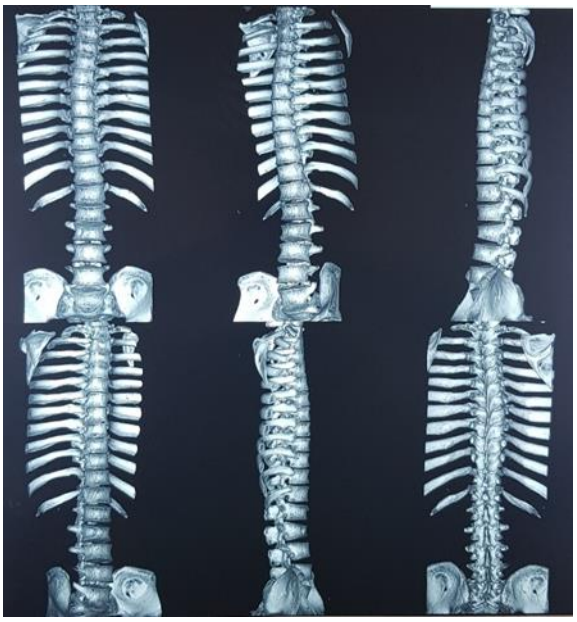

Figure 3. Whole spine CT-Scan showing thoracal scoliosis with wedge fracture at L2 level

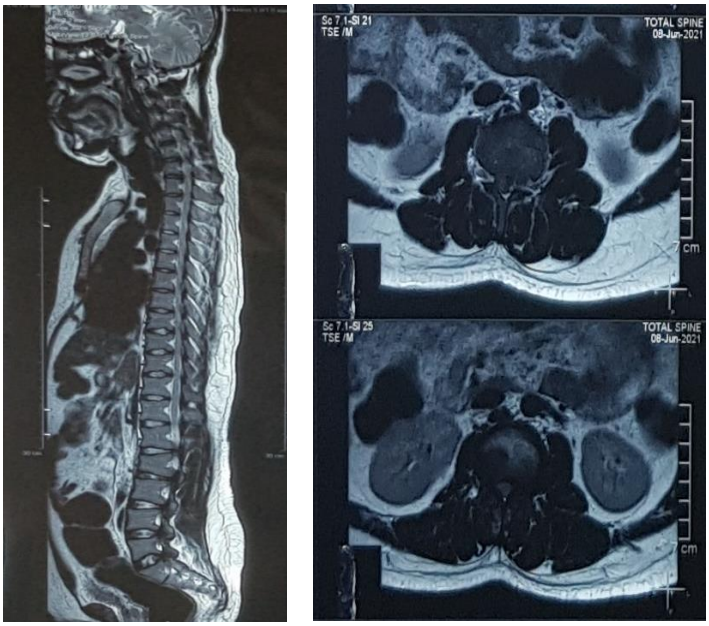

Figure 4. Whole Spine Sagittal View and Axial MRI showing stenosis in several level of lumbar spine and ossification of ligamentum flavum

At first, he admitted to the Public Hospital at his home town Tanjung Pinang, Riau accompanied by his family. He did consume some medical therapies and fisioterapies. After no improvement with his symptoms, then he referred to another Hospital at Pekanbaru, Riau.

For further scanning and intervention, in June 2021 he was referred to Public Hospital in Yogyakarta. He was scheduled to undergo surgery there, but after several consideration he decided to do examination at RSA UGM on December 2021.

At Orthopaedic Clinic RSA UGM patient came with thoracolumbar spinal orthosis, in physical examination revealed a short stature only 115 cm, 45 kg body weight, large head size with prominent forehead, nose-bridge depression, short arms and legs with long trunk and thoracal mild scoliosis. (Figure 1). By the time he came to our clinic the motoric and sensoric symptoms still persist, but the low back pain already decreased.

Radiographs revealed mild scoliosis and reduced interpediclar distance of thoracal spine. (Figure 2).
On 3D CT-Scan the thoracal scoliosis can be seen clearly with wedge fracture at L2 level of lumbar spine. (Figure 3). MRI of thoracolumbar spine revealed several spinal canal stenosis at L2-L5 level of lumbar spine accompanied by ossification of the ligamentum flavum. (Figure 4).

Three stages of laminectomy and posterior decompression at L2-L5 level followed by pedicle screw instrumentation were performed. The patient was placed in prone position, at the first we marked the incisional area with C-ARM. General anaesthesia was used during the operation. Pedicular screws were inserted by the free hand technique under C-ARM guidance from L2 to L5. Wide decompressive laminectomy with spinous process-splitting approach and resection of the Ossification of Ligamentum Flavum (OLF) were performed at L2 to L5 level using a high-speed burr and a Kerrison rongeur.

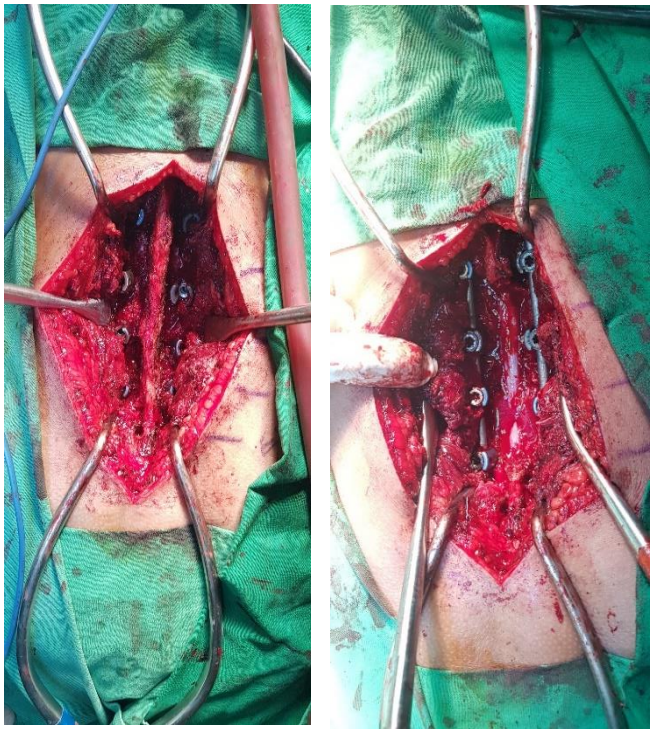

Figure 5. Insertion of pedicle screw followed by laminectomy decompression of L2-L5 level of lumbar spine and stabilization

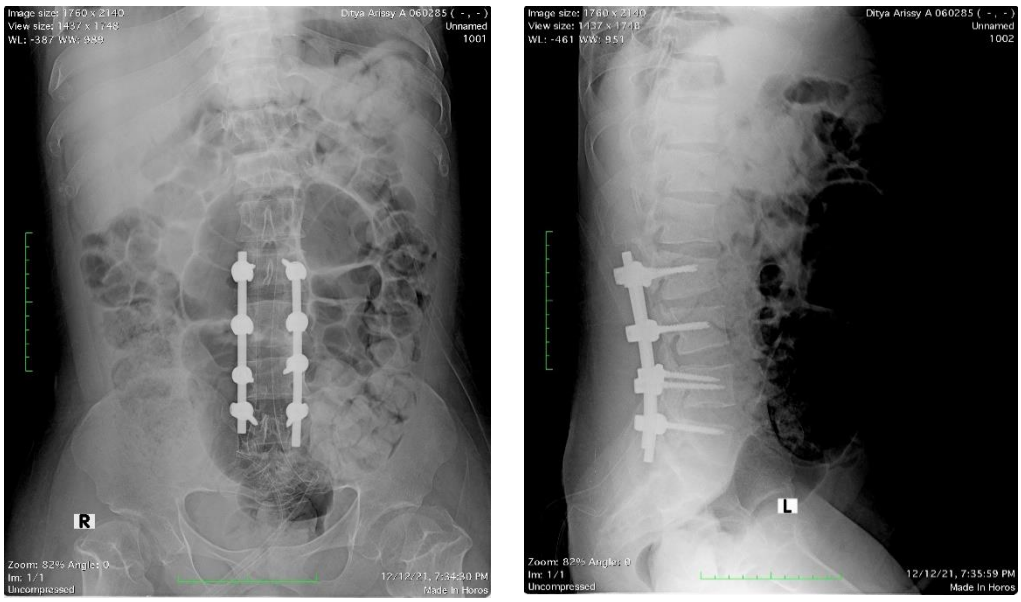

Figure 6. Post operative AP and lateral radiographic images

After surgery the patient was admitted to intensive care unit for 1 day, then moved to Orthopaedic ward afterwards to continue the post-surgery medication and do physiotherapy to encourage his movements. Plain radiograph of lumbosacral spine was taken which revealed good instrument position (Figure 6).

The patient was discharged 3 days later. Two months after the surgery, the patient's motoric function was improved significantly. He could barely stand on his own feet without holding into handrails or wall. The neurogenic claudication caused by nerve entrapment was completely dissapear. Meanwhile, the sensoric problems still persist. Physiotherapy and muscle exercise was encouraged in order to strengthen back and extremities muscles.

3. Discussion

Most of all achondroplastic patients have a disproportionally large cranial cranial vault with a bulging forehead, though hydrocephalus requiring dranage is an occational feature. Spinal symptoms with neurological complications are frequently reported, though clearly these are self-selected by the need to attend the hospital (2). The clinical expression of lumbar spinal stenosis ranges from low back pain with neurogenic claudication to paraplegia with bladder and bowel dysfunction. Achondroplasia-associated lumbar spinal stenosis results from a combination of congenital dysplasia and acquired degenerative changes. Spinal dysplasia in Achondroplasia is due to disordered endochondral ossification resulting in early fusion of the pedicles to the vertebral body. Hence the cross-sectional area of the spinal canal is narrowed by the shortened pedicles and the decreased interpedicular distance. The dimensions of the spinal canal are further reduced by age-related degenerative changes, which eventually compromise the space available for the neutral elements (3).

In patients with achondroplasia, congenital spinal stenosis is not severe enough in

itself to cause neurological impairement (4). The neurologic impairement was reported to be caused by several factors, including disc degeneration, kyphosis of lumbar vertebrae, ossification of ligamentum flavum, bony spurs, and thickened laminae and facet joints (5).

During infancy the most frequent finding related to axial the axial skeleton is a mild thoracolumbar kyphosis. This has been attributed to hypotonia and great majority of these curves recover with weight bearing and ambulation. When an upright posture and walking are achieved lumbar lordosis and prominence of the buttocks increase. In 23-30% persistent kyphosis develops and 36% of these curves becomes severe. The development of kyphosis has been associated with wedging or hypoplasia of the vertebral bodies at the thoracolumbar junction (6,7). The middle column remains intact and may act as a hinge. This results in loss of anterior height of the vertebra while the posterior height remains unchanged. As the collapsed anterior vertebrae fuse together, the spine bend forward, causing kyphotic deformity. Because the majority of damage is limited to the anterior vertebral column, the fracture usually stable and rarely associated with neurologic compromise (8). Spinal compression fractures can be insidious and may produce back pain early in the course of progressive disease (9). As we can see in this patient, he experienced back pain about 12 months before came to our clinic. When he came to our clinic the back pain already relieved and the most prominent symptoms are related to his neurologic deteroriation caused by other elements at the vertebrae.

In an achondroplastic patient, there are abnormalities of intracartilaginous ossification caused by mutation of *FGFR 3* gene. Ossification of Ligamentum Flavum (OLF) also occurs through intracartilaginous ossification, they may have some correlation, but the mechanism is still unknown. The OLF is a rare condition that is reported in the Asian and Caucasian populations. Thus, the development of OLF with aging accelerated the severity of spinal canal stenosis, resulting in progressive symptoms requiring surgery at an older age. The levels of pathology were mostly reported to be thoracic and thoracolumbar, only a few studies reported pathology in lumbar region (5,10). The most common site of OLF requiring surgery is the thoracic spine (Hanakita et al. 1990). The incidence of OLF at the lumbar region has been reported to range from 8.6% to 11.3% (11,12). Considering treatment after diagnosis, it is important to decide the optimal timing of surgical intervention for achondroplastic patients with spinal stenosis. Early surgical decompression is the key to gain better neurologic recovery and pain improvement[10]. Multi-level laminectomy is indicated if the clinical features mainly result from a narrow canal. The good long-term results of multi-laminectomy for spinal stenosis are related to the short duration of preoperative symptoms (13). Perioperative complications such as dural tear are common during multi-level laminectomy in patients with achondroplasia (14). Dural ossification is a common finding in OLF (40%) and could lead to iatrogenic tears of the dura during decompression in achondroplastic patients with spinal stenosis. The incidence of dural tears and cerebrospinal fluid leakage in OLF patients was 32% (15,16). It is recommended to use high-speed burr to gradually thin the lamina before using the Kerrison rongeur to remove the bone (10).

4. Conclusion

In this report, decompressive wide laminectomy and resection of adherent OLF followed by fusion with instrumentation were performed using the same technique as we mentioned above. No dural tear was recorded on this patient. There was an improvement on patient's neurological symptoms and function after the surgery. This method is safe and effective.

5. Acknowledgement

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