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Case Report

Achondroplasia with seronegative spondyloarthropathy resulting in recurrent spinal stenosis : A case report

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ABSTRACT

Background: Achondroplasia is an autosomal dominant condition caused by the G380 mutation of the gene encoding fibroblast growth factor receptor 3 on chromosome 4P. The classical findings include rhizomelic extremities, short stature, and spinal stenosis involving the upper cervical and distal lumbar spine. Rarely, achondroplasia coexisting with seronegative spondyloarthropathy can result in recurrent canal stenosis. Here, we report a 36-year-old male with symptomatic recurrent L3-L4 spinal stenosis 9 years following an original L2-S1 lumbar decompression for stenosis.

Case Description: A 36-year-old male with achondroplasia (height of 113 cm and weight 43 kg [BMI-33.7]) presented with low back and right lower extremity sciatica (ODI 39). He had achondroplasia with a short stature. Nine years ago, he had an L2-S1 laminectomy for decompression of stenosis. When the new MRI revealed recurrent severe L3-4 stenosis, he underwent a repeated L3-L4 decompression with fusion. One year later, the patient was neurologically intact with radiographic confirmation of adequate L3-L4 arthrodesis.

Conclusion: A 36-year-old male with achondroplasia and a history 9 years ago of an L2-S1 laminectomy for stenosis, presented with symptoms and signs of recurrent L3-L4 stenosis that responded to repeated decompression and fusion.

Keywords: Achondroplasia, Lumbar canal stenosis, Resurgery, Seronegative spondyloarthritis, VAS score

INTRODUCTION

Achondroplasia an autosomal dominant caused by a mutation of the G380 gene encoding fibroblast growth factor receptor 3 on chromosome 4P.[1] The clinical findings include rhizomelic extremities (humerus shorter than forearm and femur shorter than tibia), cranial frontal bossing, and often cervical, thoracic, and lumbar spinal stenosis with attendant cord compression.[3] Degenerative changes at these levels typically occur at an earlier age and include disc herniations, degenerative spondylosis, and other arthritic findings. Here, a 36-year-old male with achondroplasia and seronegative spondyloarthropathy presented with recurrent L3-L4 lumbar canal restenosis 9 years following an initial L2-S1 laminectomy for stenosis.

CASE PRESENTATION

A 36-year-old male with achondroplasia (113 cm and weight 43 kg [BMI-33.7] with forehead prominence) presented with low back pain and right lower extremity sciatica of 6 months

duration. On examination, the patient was wheelchair bound, had difficulty standing, and could not walk independently (4/5 motor dysfunction diffusely throughout both lower extremities and diffuse hypoesthesia in the L3, L4 distributions). Nine years ago, he had undergone a L2-S1 decompressive laminectomy for stenosis with good resolution of his symptoms. New radiographic studies now showed recurrent L3-L4 lumbar stenosis. Plain lumbar X-rays demonstrated fusion between the L1-L3 and L3-L5 lumbar levels, with osseous fusion of the right sacroiliac joint accompanied by irregularity of the left S1 joint cortex due to advanced seronegative spondyloarthropathy [Figures 1-4].

Surgery

The patient underwent a wide bilateral microscope-assisted decompressive L3-L4 laminectomy for resection of L3-L4 bony spurs, L3-4 discectomy, interbody fusion using local autograft, and L3-L4 right-sided instrumented stabilization [Table 1]. At surgery, it was difficult to identify the pedicles of L3-4 due to the distorted anatomy, hypertrophic scar, and dense fibrous bridges between the respective facet joints.

Postoperative outcomes

Postoperatively, the patient neurologically improved; he walked with a walker on the day of surgery and was able to return to work 6 weeks later. One year postoperatively, his motor deficit fully resolved (i.e. to 5/5) [Table 2].

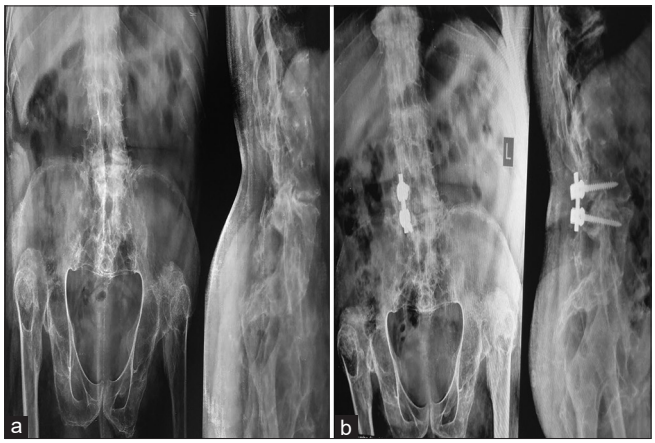

Figure 1: Preoperative X-ray of lumbosacral spine (anteroposterior and lateral): X-ray suggestive of degenerative changes of lumbar spine and osseous fusion of the right SI joint with irregularity of cortex in the left SI joint. (b) Postoperative X-ray lumbosacral spine (anteroposterior and lateral): posterior decompression at L3-4 and fusion was achieved with the right side unilateral instrumented stabilization and interbody fusion with bone graft.

DISCUSSION

Patients with achondroplasia and congenitally shortened pedicles are susceptible to developing cervical, thoracic, and/

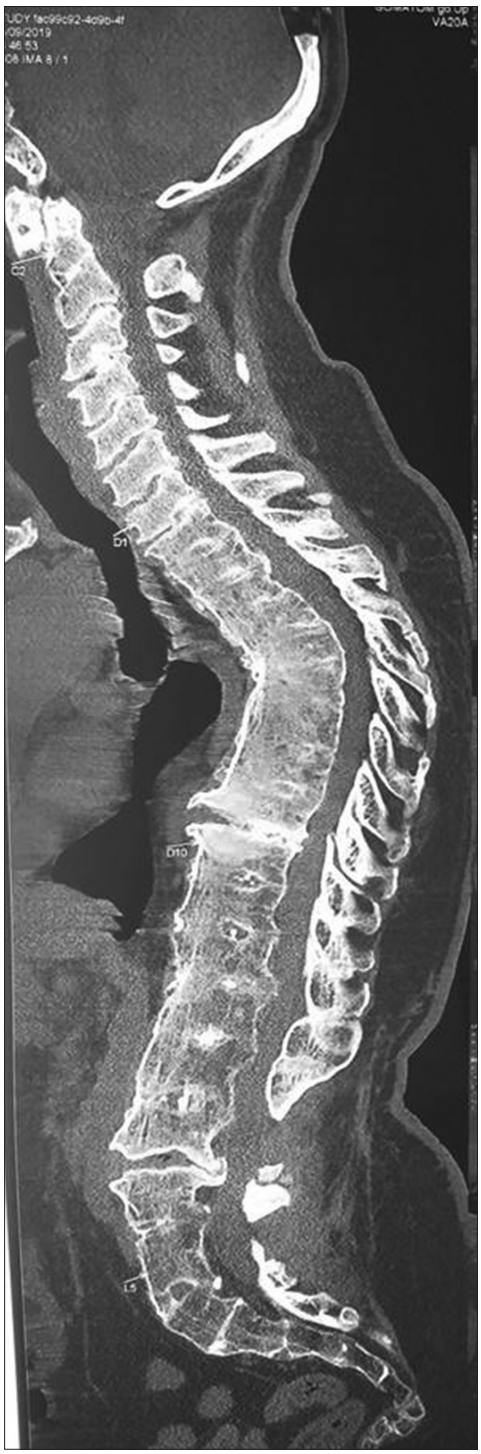

Figure 2: CT scan sagittal view: L3-4 unfused segment with fused dorsolumbar segments showing advanced seronegative changes.

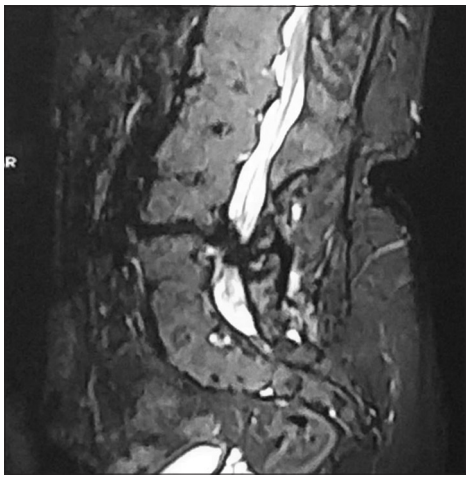

Figure 3: MRI scan of lumbosacral spine sagittal view: lumbar canal stenosis is seen at L3-4 level due to diffuse disc bulge, facetal arthropathy, and osseous malformation of L3 and L4 vertebral bodies.

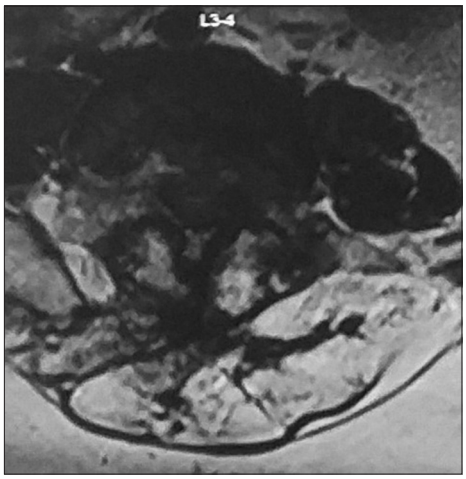

Figure 4: MRI scan of lumbosacral spine axial view: bilateral L3-4 foraminal narrowing and lumbar canal stenosis are seen causing bilateral L3 nerve root compression.

or lumbar stenosis. Patients typically become symptomatic in their 30s though 50s due to accelerated disc degeneration, lumbar kyphosis, hypertrophy of ligamentum flavum, bone spurs, and thickened laminae/facet joints.[2,5]

There is often a need for revision spine surgery in these patients attributed to their accelerated facet hypertrophy associated with their genetic defect (i.e., an exaggerated response to normal motion leading to early degeneration)[4,6] [Table 3]. Ain *et al*. further reported recurrent stenosis occurring in these as well.[1] Further, repeat surgery may successfully reduce pain and neurological symptoms/signs. In the case presented of a 36-year-old male with achondroplasia, 9 years following an original L2-S1 laminectomy for stenosis, repeated decompression and fusion at the L3-L4 level addressing recurrent stenosis were successful.

Table 1: Surgical outcomes.

Surgical outcomes	Duration of surgery	Blood loss	Hospital stay	Complications
	3 h	200 ml	3 days	None

Table 2: Patient scores.

	VAS score	ODI score	Motor power of L3, L4
Preoperative findings	8/10	39	4/5
One year postoperative findings	1/10	4	5/5

Table 3: Review of revision spine surgeries in achondroplasia.

Revision spine surgery in achondroplasia	Number of cases	Mean interval between most recent revision surgeries
Pyeritz *et al*.[4]	9	3.3 years (range 3 months –9.5 years)
Ain *et al*.[1]	8	8.2 years (range 2–18 years)

CONCLUSION

Patients with achondroplasia who have previously undergone lumbar decompressive surgery may develop recurrent lumbar stenosis that responds well to repeated surgical intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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