

CASE REPORT

Open Access



Double non-contiguous fractures in a patient with spondylo-epiphyseal dysplasia with spinal ankylosis treated with open and percutaneous spinal fixation technique: a case report

Takahiro Ushijima^{1,2}, Kenichi Kawaguchi^{1,3}, Tadashi Matsumoto⁴, Masaki Takagi⁵, Tatsuro Kondoh⁴, Gen Nishimura⁶, Aritoshi Iida⁷, Shiro Ikegawa⁷, Nobuhiko Haga⁸ and Go Kato^{1,2*}

Abstract

Background: Patients with ankylosing spines are susceptible to developing spinal fractures even with minor trauma and can develop early or late neurological injuries. These fractures require early and aggressive surgical management to enable spinal stability and/or neural decompression. Being highly unstable by nature, they require relatively long segment instrumentation and fusion, which can increase paravertebral soft tissue damage and perioperative bleeding. The purpose of this report is to describe a rare case of traumatic double fractures at the cervico-thoracic and thoraco-lumbar transition zones in ankylosing spine with spondylo-epiphyseal dysplasia (SED) of unknown cause, which were successfully treated with a combined open and percutaneous spinal fusion procedure.

Case presentation: A 46-year-old woman who was diagnosed with non-contiguous fractures in cervico-thoracic and thoraco-lumbar junction zones among multiple injuries sustained in a traffic accident was treated with hybrid techniques for posterior instrumentation with an open approach using a computed tomography (CT)-based navigation system and percutaneous pedicle-screwing method. She regained mobility to pre-admission levels and started walking on crutches 3 months postoperatively. Genetic testing for the cause of SED revealed no mutation in the COL2A1 or TRPV4 genes. The union of fractured spine was confirmed on CT scan 1 year postoperatively.

Conclusion: This is the first report of double spinal fractures in an ankylosing spine with genetically undetermined spondyloepiphyseal dysplasia. A long-segment posterior instrumentation procedure incorporating the invasive treatment of spinal fractures in ankylosing spondylitis or diffuse idiopathic hyperostosis was effective.

Keywords: Trauma, Spine, Spondylo-epiphyseal dysplasia, Ankylosing spine, Spinal fracture

*Correspondence: gkato23@yahoo.co.jp

¹ Department of Spine Surgery, Saga Medical Centre, Koseikan, 400 Nakabaru Kase-Machi, Saga 840-8571, Japan

Full list of author information is available at the end of the article

Background

Ankylosing spondylitis (AS) or diffuse idiopathic skeletal hyperostosis (DISH) commonly accompanies ankylosing spines; spinal fractures frequently occur with minor trauma [1, 2] and are extremely unstable because of the long lever arms of the fused spinal column and several complications, including common early and late neurological symptoms [2]. Therefore, early surgery for neural decompression and spinal stability is recommended [3, 4]. A case with double fractures of an ankylosing spine with genetically undetermined spondyloepiphyseal dysplasia (SED) and spinal ankylosis was successfully treated with hybrid open and percutaneous spinal fusion.

Case presentation

A 46-year-old woman involved in a traffic accident was brought to our hospital. On arrival at the emergency room, her vital signs were stable and physical examination revealed no neurological deficit. Whole body computed tomography (CT) confirmed double spinal fractures at the cervicothoracic (C-T) and thoracolumbar (T-L) junctional zones in an ankylosing spine, with atlas to coccyx fusion, as well as traumatic hemopneumothorax and multiple rib fractures. A right thoracic curve with a Cobb angle of 15°, a sagittal kyphotic thoracic curve with a Cobb angle of 73°, sacral anteversion, and coccygeal retroversion were observed (Fig. 1). Magnetic resonance imaging of the spine revealed a mild dural sac indentation at the T-L junctional zone, but without spinal cord compression.

In infancy, the patient had progressive multiple joint contractures indicating arthrogyryposis multiplex congenital, which was not genetically confirmed. Height and weight on admission were 130 cm and 40 kg, respectively. No visual or acoustic deficits were evident. Whole body roentgenograms revealed marked osteoarthritic changes in almost all joints and ankyloses in the knees and shoulders. She had no medical history of fractures and underwent left and right hip joint replacement surgeries at 32 and 39 years of age, respectively (Fig. 2). Respiratory distress from traumatic hemopneumothorax resolved within 8 days of hospitalization, and the patient subsequently underwent surgical posterior spinal fusion of both spinal fractures without bone grafting (Fig. 3).

The patient underwent tracheal intubation in the neutral cervical spine position using a monitor-integrated video laryngoscopy. She was then placed in the prone position taking care not to displace floating thoracic spine fragments. Spinal alignment in the prone position was checked using fluoroscopy and CT; a mattress was inserted between the body and four-post spine frame to equalize the space created by the thoracic spine kyphotic deformity, and C2–T4 and T9–L3 posterior fusion was

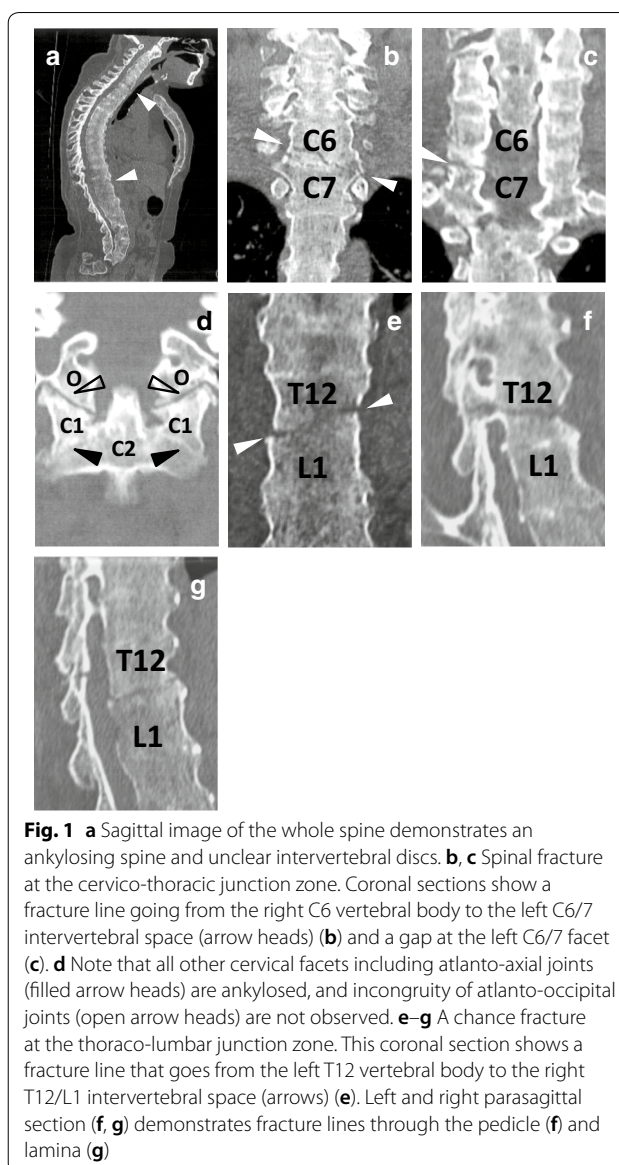
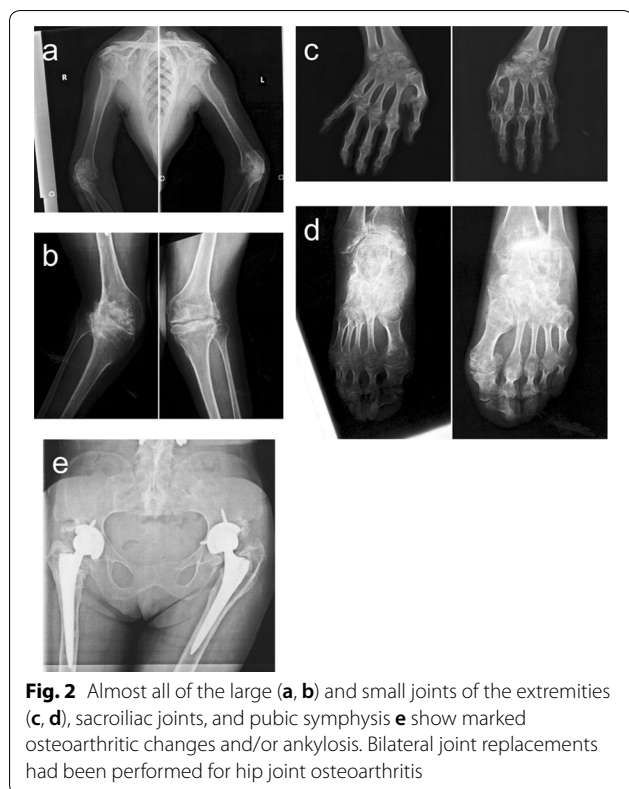


Fig. 1 **a** Sagittal image of the whole spine demonstrates an ankylosing spine and unclear intervertebral discs. **b, c** Spinal fracture at the cervico-thoracic junction zone. Coronal sections show a fracture line going from the right C6 vertebral body to the left C6/7 intervertebral space (arrow heads) (**b**) and a gap at the left C6/7 facet (**c**). **d** Note that all other cervical facets including atlanto-axial joints (filled arrow heads) are ankylosed, and incongruity of atlanto-occipital joints (open arrow heads) are not observed. **e–g** A chance fracture at the thoraco-lumbar junction zone. This coronal section shows a fracture line that goes from the left T12 vertebral body to the right T12/L1 intervertebral space (arrows) (**e**). Left and right parasagittal section (**f, g**) demonstrates fracture lines through the pedicle (**f**) and lamina (**g**)

performed. A C2 lamina screw (left side), pedicle screw (right side), cervical lateral mass screws (C3–C6), and thoracic pedicle screws (T2–T4) were inserted via an open approach. A C-arm three-dimensional navigation system (Brainlab Spinal Navigation, Munich, Germany) was used for placing C2 and thoracic pedicle screws; lateral mass screws were placed using a lateral fluoroscope. Contoured rods from C2 to T4 and a cross-link at the T1 level were placed. Pedicle screws were percutaneously placed from L1 to L3 using biplanar fluoroscopy.

Although identifying pedicle contours of T9–T11 was difficult because of spinal osteoporosis, screw placement was safely performed without a navigation system via the open approach because the diameters of the pedicles at these levels were large enough (> 6.0 mm on the preoperative CT



scan). Pedicle screws were not placed along the left side of T9 because the pedicle was sclerosed and a rib fracture dislocation occurred while preparing the pilot hole. The 5.5-mm contoured rods were inserted through polyaxial heads of pedicle screws at the open thoracic incision and caudally passed subcutaneously through polyaxial heads of lumbar pedicle screws. Cross-links were placed between T12 and L1. Decompressive laminectomy or laminotomy was not performed because of the absence of cord compression. Surgical duration was 8 h, with approximately 1100 mL blood loss. The patient recovered well without postoperative complications and was discharged for rehabilitation 2 months postoperatively. She could walk and resume work, respectively, at 3 and 6 months postoperatively. One-year follow-up CT revealed a bony fusion of C-T and T-L fractures (Fig. 4). We suspected SED based on her medical history and whole body roentgenograms, but did not find *COL2A1* and *TRPV4* mutations; further genetic testing was not performed because no candidate gene for this phenotype was identified.

Discussion and conclusions

We report double noncontiguous spinal fractures in a patient with SED having spinal ankylosis. No similar cases have been reported, which necessitated adaptation

of spinal fracture treatments in patients with AS or DISH.

1. Fracture characteristics

AS or DISH usually accompany ankylosing spines. Non-contiguous spinal fractures in patients with ankylosing spine, as in the present case, are not rare [1, 5]. Because of poor bone quality, the spine is brittle, osteoporotic, and stiff [4, 6, 7], increasing susceptibility to fracture even with low-energy trauma [8, 9]. Fracture pattern in patients with AS typically involves three columns [10], as in this case. Compared with previous reports, this study is unique because the fracture occurred in an ankylosing spine with SED. The cervical spinal fracture in our patient was observed at the C6–C7 segmental level, consistent with previous reports [1, 4, 11]. Spinal fractures in patients with AS frequently accompany hyperextension fractures [1, 11] and are classified into four patterns based on fracture excursion through intervertebral disc, vertebral body, or both [1]. However, coronal CT in our case demonstrated a gap between the left and right fracture lines at both cervical and thoracic vertebral bodies—a pattern not previously observed. Based on the accident and the difference of number of left and right rib fractures (4:1), we attribute the pattern of fracture lines to direct or indirect lateral external forces impacting the spine.

2. Etiology of ankylosing spine

Congenital SED is a rare form of skeletal systemic disease [12]. Previous studies have demonstrated that patients with congenital SED present with a variety of deformities in the spine, including instability of the atlanto-axial joint, progressive kyphoscoliosis, platyspondylitis, lordotic lumbar vertebrae, and pear-like shape to the corps vertebrae at the thoracolumbar interface [13, 14]. However, our patient had extremely rare features in the spine such as ankylosing spine and the absence of instability of the atlanto-axial joint with SED. To our knowledge, only two studies have reported ankylosing spines with SED [15, 16], wherein patients exhibited X-linked recessive inheritance and associated mutations of transport protein particle *TRPPC2* [17], with onset later than congenital SED. This patient probably had another causative gene because of her sex and appearance of spinal ankylosing and multiple joint contractures in infancy. Because SED is a skeletal disorder mediated by *COL2A1*, we investigated for gene mutations [18] but found none. Moreover, no mutation of *TRPV4*, reportedly related to spondylometaphyseal dysplasia (Kozłowski type) and SED (Maroteaux type) [19], was observed. The genetic etiopathology in our patient remains unknown.

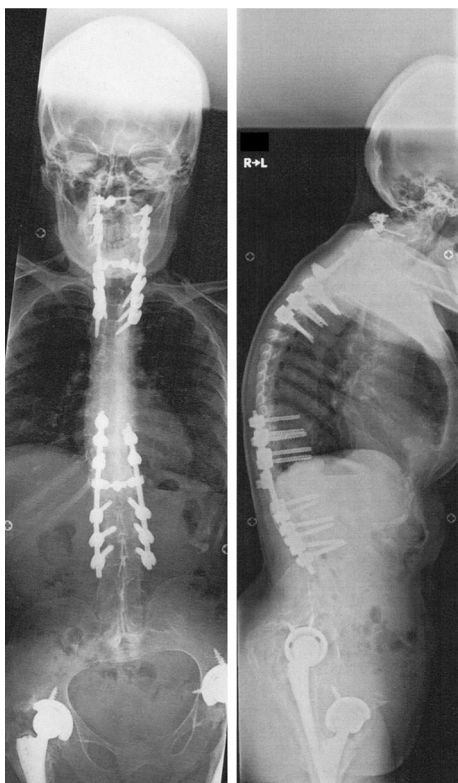


Fig. 3 The anteroposterior (left) and lateral (right) roentgenograms of spinal fusion of both fractures at the cervico-thoracic and thoraco-lumbar junctions without bone grafting



Fig. 4 Sagittal computed tomography image of the whole spine 1 year postoperatively

3. General problems treating ankylosing spinal fractures

Ankylosing spine creates high instability at fracture site because of long lever arms secondary to spinal column stiffness. Fractures frequently occur at the C-T and T-L transitional zones, which are subjected to extension force when patients are supine, resulting in delayed union [20, 21], epidural hemorrhage, and/or late-onset paralysis [22, 23]. Furthermore, a much higher mortality rate is reported in patients with cervical spinal fracture who have AS (> 30%) [1, 11] than in those without AS (18%) [11]. Therefore, early aggressive surgeries with posterior and/or anterior fixation are recommended. A disadvantage of treating this fracture type is the increase in the number of fused spinal segments (average, 5.6 segments) [4]; it is generally recommended to make multiple anchor points by extending the instrumentation over at least three vertebral levels above and below the fracture site [4, 24].

4. Specific devices used for treatment

Our patient had extensive thoracic kyphosis; therefore, we carefully monitored motor- and

somatosensory-evoked potentials intraoperatively. To prevent intraoperative worsening of spinal alignment in the prone position, secondary to contact pressure disparities at the frontal truncal surface due to the rigid and kyphotic spine, we inserted cushioning materials into the space between the trunk and operating table. A CT-based navigation system was used for placing pedicle screws at C2 and the thoracic spine, considering the extreme difficulty in identifying anatomical landmarks for pedicle screw insertion. We made C2 anchors because we were concerned that cervical lateral mass screws had significantly lower resistance to pull-out forces than pedicle screws [25]. No new fracture was observed in our case at the thoracic spine without instrumentation between T4 and T9 partly due to her lower activity level secondary to ankyloses of all joints and spine. However, even with similar fracture patterns, non-skip cervicolumbar fusion would be favorable in patients with higher activity levels. Eventually, bony

fusion of three-column spinal fractures at C-T and T-L junctional zones using hybrid open and percutaneous spinal fusion techniques was successful, despite difficulties ensuing from SED and spinal ankyloses of idiopathic genetic etiopathogenesis.

Abbreviations

SED: spondylo-epiphyseal dysplasia; CT: computed tomography; AS: ankylosing spondylitis; DISH: diffuse idiopathic skeletal hyperostosis; C-T: cervico-thoracic; T-L: thoraco-lumbar.

Authors' contributions

TU, KK, and GK have been involved in the clinical management, data acquisition and interpretation of data of the patient. TM, TK, GN, AI, SI and NH were involved in genetic testing for TRPV1 mutation. GN and MT performed genetic testing for COL2A1 mutation. TK, GN SI and NH were involved in image diagnosis of the patient. TU, KK and GK drafted the manuscript. All authors read and approved the final manuscript.

Author details

¹ Department of Spine Surgery, Saga Medical Centre, Koseikan, 400 Nakabaru Kase-Machi, Saga 840-8571, Japan. ² Department of Trauma Centre, Saga Medical Centre, Koseikan, 400 Nakabaru Kase-Machi, Saga 840-8571, Japan. ³ Department of Orthopedic Surgery, Kyushu University Graduate School of Medical Sciences, 3-1-1 Maidashi Higashi-Ku, Fukuoka 812-8582, Japan. ⁴ Division of Developmental Disability, Misakaenosono Mutsumi Developmental Medical and Welfare Center, 570-1 Konagaichomaki, Isahaya 859-0164, Japan. ⁵ Department of Endocrinology and Metabolism, Tokyo Metropolitan Children's Medical Center, 2-8-29 Musashidai Fuchu, Tokyo 183-8561, Japan. ⁶ Department of Radiology, Tokyo Metropolitan Children's Medical Center, 2-8-29 Musashidai Fuchu, Tokyo 183-8561, Japan. ⁷ Laboratory of Bone and Joint Diseases, RIKEN Center for Integrative Medical Sciences, 4-6-1 Shirogane Dai Minato-Ku, Tokyo 108-8639, Japan. ⁸ Department of Rehabilitation Medicine, Graduate School of Medicine, The University of Tokyo, 7-3-1 Hongo Bunkyo-Ku, Tokyo 113-8655, Japan.

Acknowledgements

Not applicable.

Competing interests

The authors declare that they have no competing interests.

Availability of data and materials

The datasets used during the current report are available from the corresponding author on reasonable request.

Consent for publication

The patient provided informed written consent for the publication of her data.

Ethics approval and consent to participate

Not applicable.

Funding

Not applicable.

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Received: 10 October 2017 Accepted: 1 February 2018

Published online: 07 February 2018

References

- Caron T, Bransford R, Nguyen Q, Agel J, Chapman J, Bellabarba C. Spine fractures in patients with ankylosing spinal disorders. *Spine (Phila Pa 1976)*. 2010;35(11):E458–64.

- Osgood C, Abbasy M, Mathews T. Multiple spine fractures in ankylosing spondylitis. *J Trauma*. 1975;15(2):163–6.
- Broom MJ, Raycroft JF. Complications of fractures of the cervical spine in ankylosing spondylitis. *Spine (Philla Pa 1976)*. 1988;13(7):763–6.
- Kanter AS, Wang MY, Mummaneni PV. A treatment algorithm for the management of cervical spine fractures and deformity in patients with ankylosing spondylitis. *Neurosurg Focus*. 2008;24(1):E11.
- Olerud C, Frost A, Bring J. Spinal fractures in patients with ankylosing spondylitis. *Eur Spine J*. 1996;5:51–5.
- Ball J. Enthesopathy of rheumatoid and ankylosing spondylitis. *Ann Rheum Dis*. 1971;30(3):213–23.
- Carette S, Graham D, Little H, Rubenstein J, Rosen P. The natural disease course of ankylosing spondylitis. *Arthritis Rheum*. 1983;26(2):186–90.
- Einsiedel T, Schmelz A, Arand M, Wilke HJ, Gebhard F, Hartwig E, Kramer M, Neugebauer R, Kinzl L, Schultheiss M. Injuries of the cervical spine in patients with ankylosing spondylitis: experience at two trauma centers. *J Neurosurg Spine*. 2006;5(1):33–45.
- Thumbkat P, Hariharan RP, Ravichandran G, McClelland MR, Mathew KM. Spinal cord injury in patients with ankylosing spondylitis: a 10-year review. *Spine (Philla Pa 1976)*. 2007;32(26):2989–95.
- El Teclé NE, Abode-lyamah KO, Hitchon PW, Dahdaleh NS. Management of spinal fractures in patients with ankylosing spondylitis. *Clin Neurol Neurosurg*. 2015;139:177–82.
- Murray GC, Persellin RH. Cervical fracture complicating ankylosing spondylitis: a report of eight cases and review of the literature. *Am J Med*. 1981;70(5):1033–41.
- Spranger JW, Wiedemann HR. Dysplasia spondyloepiphysaria congeinta. *Helv Paediatr Acta*. 1966;21:598–611.
- Hensinger RN, Ewen GDM. Congenital anomalies of the spine. In: Rothman RH, Simoeone FA, editors. *The spine*. 2nd ed. Philadelphia: WB Saunders; 1982. p. 299–304.
- Tolo VT. Spinal deformity in skeletal dysplasia. In: Weinstein SL, editor. *The pediatric spine*, vol. 1. New York: Raven Press; 1994. p. 369–96.
- Bos J, Rogge CW. A family with late spondyloepiphyseal dysplasia. *Ned Tijdschr Geneesk*. 1974;118(16):576–81.
- Job-Deslandre C, Menkès CJ. Spondylo-epiphyseal dysplasia with ankylosing development. Apropos of a case. *Rev Rheum Mal Osteoartic*. 1991;58(9):635–6.
- Savarirayan R, Thompson E, Gécz J. Spondyloepiphyseal dysplasia tarda (SEDL, MIM #313400). *Eur J Hum Genet*. 2003;11(9):639–42.
- Spranger J, Winterpacht A, Zabel B. The type II collagenopathies: a spectrum of chondrodysplasias. *Eur J Pediatr*. 1994;153(2):56–65.
- Nishimura G, Lausch E, Savarirayan R, Shiba M, Spranger J, Zabel B, Ikegawa S, Superti-Furga A, Unger S. TRPV4-associated skeletal dysplasias. *Am J Med Genet C Semin Med Genet*. 2012;160C(3):190–204.
- Deburge A, Guigui P, Ouahes M, Barre E. Cervical pseudoarthrosis in ankylosing spondylitis. A case report. *Spine (Phila Pa 1976)*. 1996;21(23):2801–5.
- Mountney J, Murphy AJ, Fowler JL. Lessons learned from cervical pseudoarthrosis in ankylosing spondylitis. *Eur Spine J*. 2005;14(7):689–93.
- Aoki Y, Yamagata M, Ikeda Y, Nakajima F, Nakajima A, Nakagawa K, Ohtori S, Inaoka T, Takahashi K. Failure of conservative treatment for thoracic spine fracture in ankylosing spondylitis: delayed neurological deficit due to spinal epidural hematoma. *Mod Rheumatol*. 2013;23(5):1008–12.
- Elgafy H, Bransford RJ, Chapman JR. Epidural hematoma associated with occult fracture in ankylosing spondylitis patient: a case report and review of the literature. *J Spinal Disord Tech*. 2011;24(7):469–73.
- Charles YP, Buy X, Gangi A, Steib JP. Fracture in ankylosing spondylitis after minor trauma: radiological pitfalls and treatment by percutaneous instrumentation. A case report. *Orthop Traumatol Surg Res*. 2013;99(1):115–9.
- Jones EL, Heller JG, Silcox DH, Hutton WC. Cervical pedicle screws versus lateral mass screws. Anatomic feasibility and biomechanical comparison. *Spine*. 1997;22(9):977–82.