



## Case Report

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# Chin on Chest Deformity Caused by Upper Cervical Kyphosis Associated With Ankylosing Spondylitis: A Case Report

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Chin on chest deformity caused by upper cervical kyphosis associated with ankylosing spondylitis is rare. A 66-year-old woman presented at our institute with chief complaints of difficulty in horizontal gaze and opening her mouth. Cervical radiographs showed a C0–2 angle of 1° on flexion and 7° on extension, and her chin-brow vertical angle was 49°. We planned fixation surgery at C0–5 posteriorly to prevent the progression of kyphosis, with slight correction of the kyphosis at C0–2. The correction was performed by pushing down the overlordotically contoured titanium rods connected to an occipital plate onto the C3–5 lateral mass screws, just like cantilever technique. No palpable cracking or loss of resistance was noticed during the correction. However, intraoperative radiographs revealed apparent anterior separation of the vertebral bodies between C3 and C4. Postoperative computed tomography images at the C3/4 level suggested hemorrhage from the fracture site. Tracheostomy was performed because of massive edema around the pharynx. To secure solid bone fusion, staged surgery to extend the fusion to T3 and to graft an additional iliac bone was performed. Fortunately, the C2–7 angle was corrected to 40°, and her chin-brow vertical angle was restored to 17° without any catastrophic complications. Although the patient finally obtained an ideal sagittal alignment, the surgeon should be aware that the technique had a higher perioperative risk for iatrogenic fracture, resulting in neurological and vascular injuries.

**Keywords:** Ankylosing spondylitis, Upper cervical spine, Iatrogenic fracture, Correction surgery, Kyphosis



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## INTRODUCTION

Ankylosing spondylitis (AS) is a progressive systemic type of seronegative chronic inflammatory rheumatic disease that mainly causes arthritis of the spine and sacroiliac joints and finally results in contracture or bony fusion of these joints. The symptoms usually start with back pain, followed by restricted trunk motion as the disease progresses. In severely affected patients, the entire spine, including the pelvis, becomes fixed in a flexed position.<sup>1</sup> This fixed flexion deformity causes a variety of char-

acteristic trunk dysfunction manifestations, including inability to look forward, upward, or around and difficulty in maintaining prolonged standing and supine positions.

Recently, tumor necrosis factor inhibitors have been reported to be effective in more than 60% of patients with AS, but there has been no curative treatment.<sup>1</sup> For patients who develop severe fixed flexion deformity, extensive corrective surgery of the spine is the only treatment. Surgical treatment of the thoracic and lumbar spine in patients with fixed flexion deformity had been previously reported. However, surgical treatment of the

upper cervical spine has been extremely rare.<sup>2,3</sup>

In this case report, we describe a woman who was surgically treated for chin on chest deformity caused by upper cervical kyphosis associated with AS. The patient developed iatrogenic fracture of the cervical spine during correction and eventually obtained sufficient correction because of the fracture, fortunately without any devastating complications.

## CASE REPORT

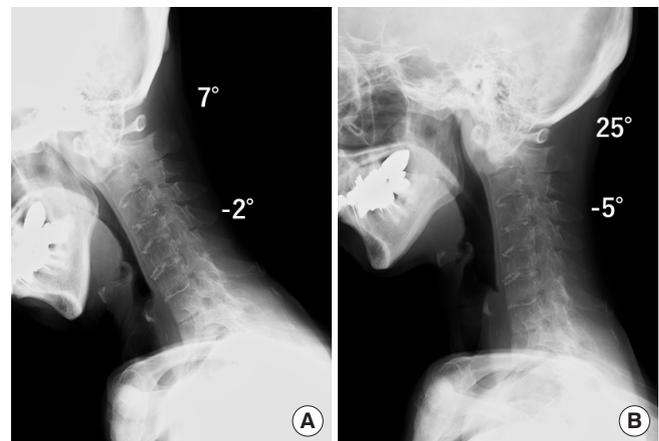
A 66-year-old woman presented to our institute with chief complaints of difficulty in horizontal gaze and opening her mouth. She started complaining of neck and shoulder pain at the age of 58 years and had been on physiotherapy and nonsteroidal anti-inflammatory drugs intake for pain control. However, at the age of 61 years, the pain worsened, and she became unable to lie down in a supine position. She had difficulty in gazing upwards at the age of 64 years. She could not keep a horizontal gaze and felt difficulty in opening her mouth because of her chin on chest position at the age of 65 years. At this point, she was diagnosed as AS at the municipal hospital and was referred to our institute for further treatment of the cervical deformity. She had no preexisting medical history, but her mother had been diagnosed as rheumatoid arthritis.

The patient's neck was fixed on flexion position and was slightly rotated to the right. She could not look in front without extending her trunk. The range of mouth opening was severely restricted to approximately 2 cm because her chest was disturbing her jaw movement. She had difficulty in taking in solid food. Her upper and lower extremities had full muscle strength but

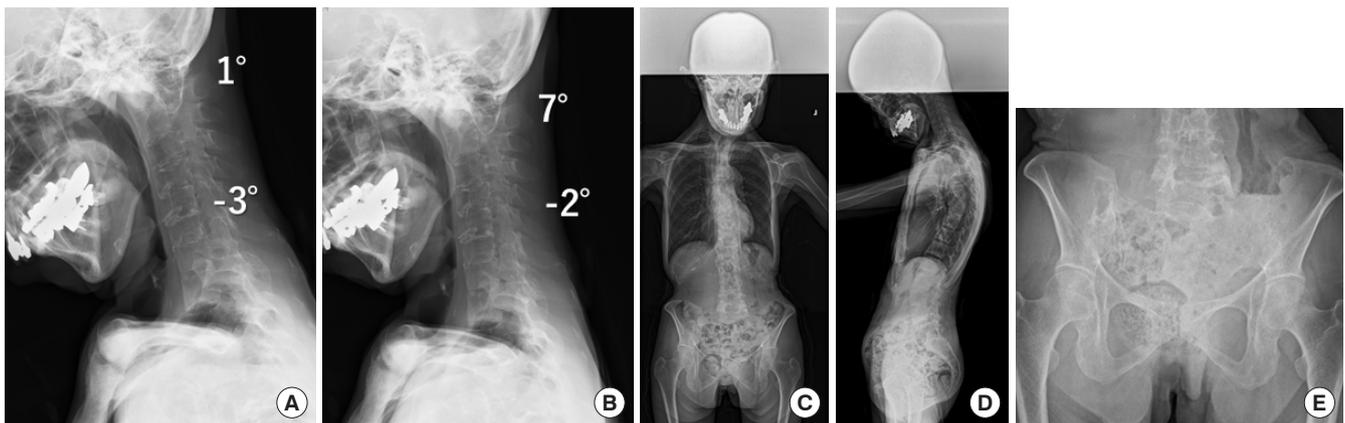
had slightly restricted range of motion. She had no apparent neurologic symptoms, based on the normal deep tendon reflexes and the absence of sensory disturbances.

A genetic test was positive for *HLA-B52* and *B62* but negative for *HLA-B27*, one of the representative genes for AS. Dual-energy x-ray absorptiometry showed osteoporosis with T score of -4.2 at L2-4 and -3.3 at the femoral neck. The Bath Ankylosing Spondylitis Disease Activity Index score was 8.4 points.

Cervical radiographs at the age of 63 years showed a C0-2 angle of 7° on flexion and 25° on extension, showing 18° of range of motion (Fig. 1). The middle to lower cervical spine was straight and fixed, with a C2-7 angle of -2°. Cervical radiographs at the age of 66 years, immediately before surgery, showed a C0-2 an-



**Fig. 1.** Cervical radiographs at the age of 63 years. Three years before surgery, the C0-2 angle is 7° on flexion (A) and 25° on extension (B). The C2-7 angle of -2° on flexion and -5° on extension indicates that the middle cervical spine is almost fixed.



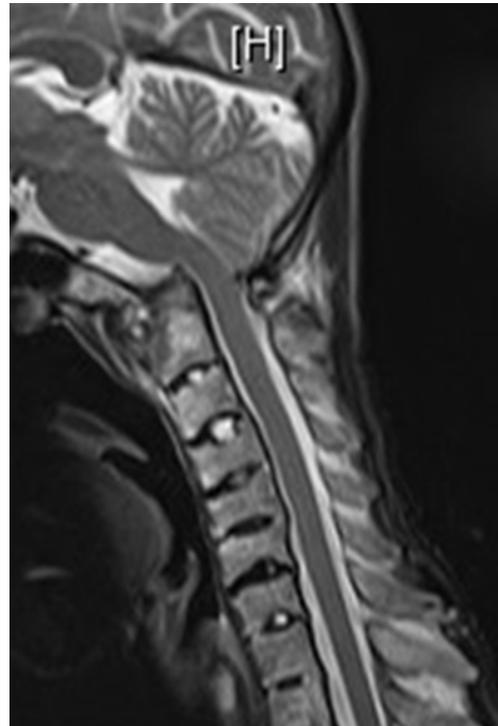
**Fig. 2.** Cervical radiographs at the age of 66 years. Immediately before surgery, the C0-2 angle of 1° on flexion (A) and 7° on extension (B) indicates a slight motion between C0 and C2. The patient is coronally and sagittally balanced (C), but her chin-brow vertical angle is 49° (D). (E) A radiograph of the sacroiliac joints showed ankylosis on the left.

gle of  $1^\circ$  on flexion and  $7^\circ$  on extension, indicating progression of the C0–2 kyphosis for 2 years (Fig. 2). A radiograph of the sacroiliac joints showed ankylosis on the left (grade 3) (Fig. 2E). Her chin-brow vertical angle was  $49^\circ$ . Sagittal images on preoperative computed tomography (CT) (Fig. 3) and magnetic resonance imaging (Fig. 4) showed basilar impression and mild compression of the medulla oblongata. However, neurological examination revealed that she was neurologically intact. Contrast CT revealed ponticulus posticus on the right (Fig. 3).

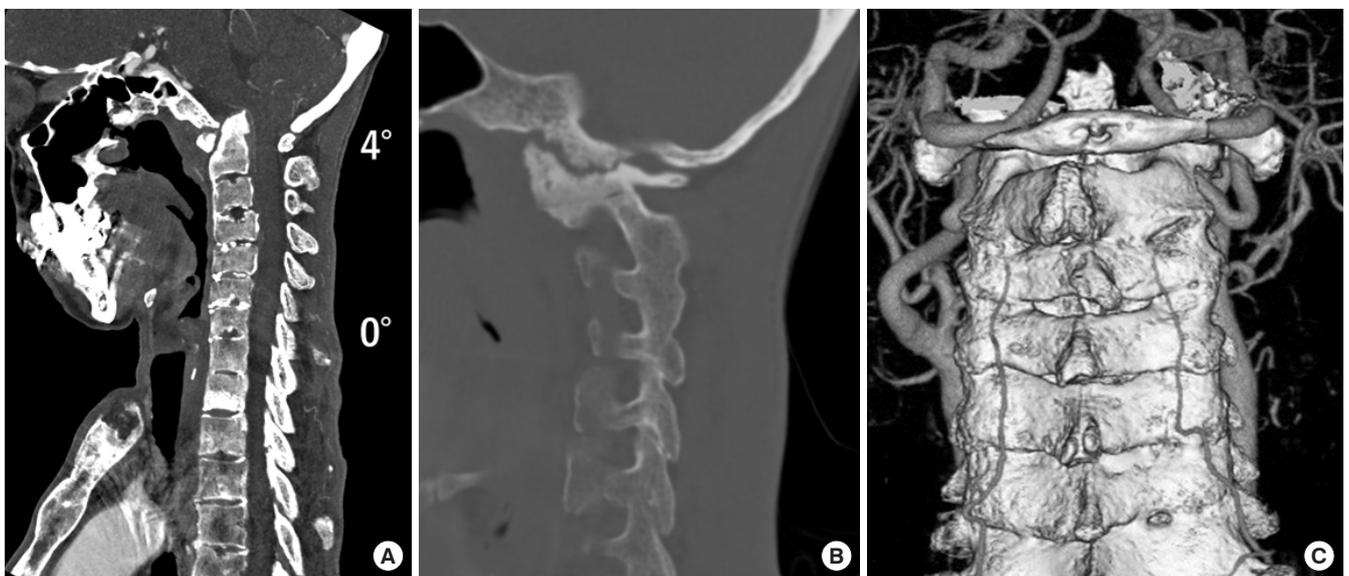
Although 3-column osteotomy at the upper or middle cervical spine would have been effective in obtaining adequate correction, it was technically highly demanding and was expected to have high risks for perioperative neurological and vascular complications. In addition, C0–1 seemed to be mobile and could expect a slight correction, which might slightly improve the chin on chest deformity. Considering the safety, we planned fixation surgery at C0–5 posteriorly to prevent kyphosis progression, with slight correction of kyphosis at C0–2.

For general anesthesia, fiberoptic visualization was used to facilitate nasotracheal tube insertion to secure airway access. The patient was placed in a prone position with 2 kg of horizontal traction to stabilize the head position. The occipital plate was placed, followed by placement of the pedicle screw only on the right side of C2. A lamina screw was placed on the right side of C2, because of the high riding vertebral artery on the left side. Lateral mass screws were placed on both sides of C3, C4, and C5. Thereafter, over lordotically contoured titanium rods

with a diameter of 3.5 mm were fixed on both sides of the occipital plate, and the correction was performed by gradually pushing down the bilateral rods onto the C3–5 screws using the



**Fig. 4.** Sagittal image on magnetic resonance imaging. There are basilar impression and mild compression of the medulla oblongata.



**Fig. 3.** Preoperative computed tomography (CT) images. (A) The sagittal images show basilar impression. (B) The C0/1 seems mobile, whereas the rest of the segments seem fused. (C) Contrast CT reveals ponticulus posticus on the right side.

cantilever technique. The rods were contoured to create lordosis along C0–2. We did not perform any osteotomies to the posterior elements for the kyphosis correction. Although there was resistance from the rod rigidity, we felt neither palpable clacking nor any apparent motions at the facet joints at C3/4.

An intraoperative radiograph after fixing the rods on the C3–5 screws revealed that the C2–7 angle was corrected to 28°. However, there was apparent anterior opening of the vertebral bodies between C3 and C4 (Fig. 5). There were no changes in the vital signs, including blood pressure and pulse rate, and no ab-

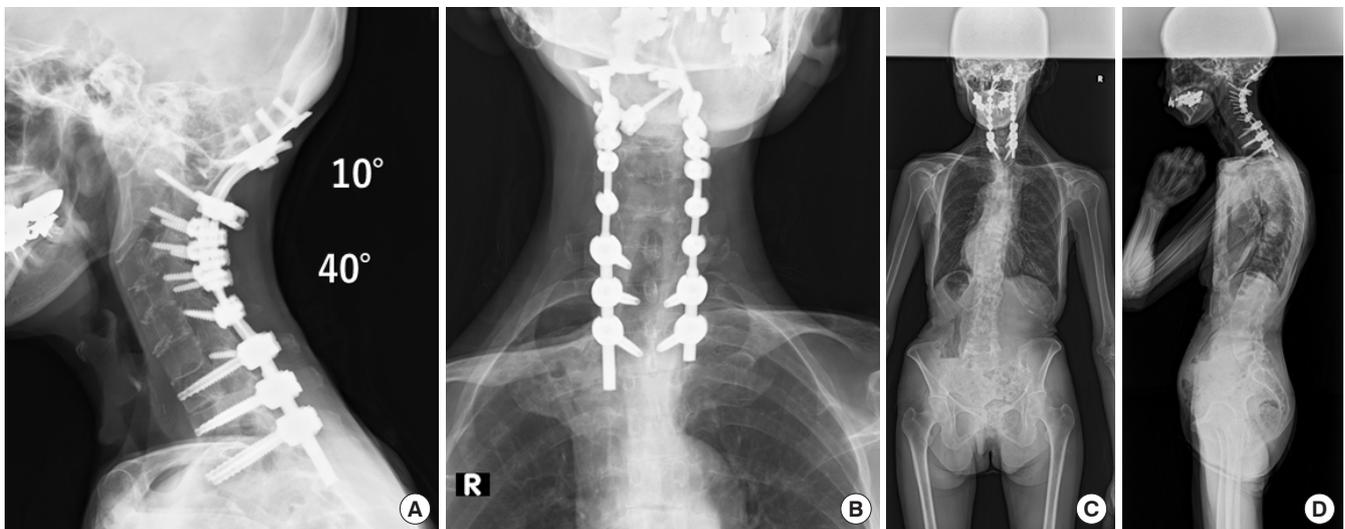
normality in the motor evoked potential responses. Eventually, the surgery was finished after local bone grafting.

After surgery, the patient's consciousness and limb movements appeared to be normal, even while intubated. Notably, the postoperative CT images showed massive swelling anterior to the C3/4 level; this suggested hemorrhage from the fracture site (Fig. 5). Massive swelling around the pharynx made postoperative extubation difficult; therefore, tracheotomy was performed.

A solid bone fusion between the C3 and C4 was expected to take a long time, and the stability of the lateral mass screws on



**Fig. 5.** Cervical radiographs and computed tomography (CT) during and immediately after correction surgery. (A) The intraoperative radiograph shows that C2–7 angle is corrected to 28°. (B) However, there is an apparent anterior opening of the vertebral bodies between C3 and C4. (C) Postoperative CT shows massive swelling anterior to the C3/4 level; this suggests hemorrhage from the fracture site. (D) Postoperative CT at 1 month after surgery revealed that the swelling at retropharyngeal space was recovered to normal status.



**Fig. 6.** Radiographs at 2 years after surgery. (A, B) The C0–2 angle is 10° and the C2–7 angle is 40° without any correction loss. (C, D) She is coronally and sagittally balanced and her chin-brow vertical angle is corrected to 17°.

C4 and C5 with lacking anterior column support at C3/4 was considered unreliable to maintain the correction. External fixation by bracing would be another option. However, C3/4 had no anterior support, and we could not be sure that the brace would be enough to support the cervical spine until obtaining solid bone fusion, since the stability of C4 and C5 lateral mass screws were unreliable. Thus, 7 days after the first surgery, a staged surgery was performed to stabilize the C3/4 by extending the fusion to T3 to prevent postoperative complication at cervicothoracic junction, and to obtain bone fusion by grafting an additional iliac bone. One month after surgery, the swelling at retropharyngeal space was recovered (Fig. 5D), and the tracheostomy cannula was removed. At 1.5 months after surgery, videofluoroscopy revealed normal swallowing, and she was able to start food intake.

After the surgery, she was able to look in front, breathe easily, and eat solid food without difficulty. Two years after surgery, the radiograph showed C0–2 angle of 10° and C2–7 angle of 40° without any correction loss (Fig. 6). Her chin-brow vertical angle was corrected to 17°.

## DISCUSSION

The dysfunctions caused by fixed flexion deformity are usually associated with severe kyphosis at thoracic and lumbar spine levels. On the other hand, the upper cervical spine tends to be lordotic to compensate for the thoracic and lumbar kyphosis and rarely becomes kyphotic. Therefore, majority of the surgical reports had been about correction of the kyphotic thoracic and lumbar spine, and reports on correction of the upper cervical spine are extremely rare. We found only 2 cases that underwent correction surgery for upper cervical kyphosis associated with AS.<sup>2,3</sup> Lin et al.<sup>2</sup> corrected a C1/2 deformity using a technique that comprised resection of the posterior arch of C1, mobilization of the vertebral arteries, wedge osteotomy of the lateral masses in C1, and internal fixation. Grundy corrected a C1/2 deformity by a technique that comprised wedge osteotomy of the lateral masses in C1 and of the odontoid process through a posterior approach.<sup>3</sup> However, these techniques were technically highly demanding and had risks for perioperative neurologic and vascular complications.

The other option is extension open wedge osteotomy at C7/T1, which was first reported by Urist in 1958 and further elaborated by McMaster and Simmons.<sup>4–8</sup> However, in the present case, the chin on chest deformity was mainly located on the upper cervical area, and we determined that correction at C7/T1

would not have helped in treating the chin on chest deformity.

Initially, posterior correction and fusion surgery at C0–5 was planned to prevent the progression of kyphosis at C0–2 and to slightly improve the horizontal gaze impairment and restricted range of mouth opening. However, unintentional open wedge fracture occurred at the C3/4 during correction and fixation. We found 2 case reports of iatrogenic fracture in patients with AS during surgery.<sup>9</sup> Compared with the normal population with minor trauma, patients with AS are at a higher risk for spinal fracture,<sup>10–14</sup> likely because of the higher incidence of osteoporosis or osteopenia.<sup>14–16</sup> In addition, a rigid spine is unable to distribute the stress to the other segments. In the present case, the fixed segment from C0–3 formed longer lever arms, which probably added stress on the C3/4 disc level and resulted in fracture at that level, even with a regular correction maneuver with cantilever technique. We should have done fixation of C2–5 with rods before correction, and correct C0–2 by holding the head manually under the monitoring of the motion at C0–2 with fluoroscopy.

Although unintentional C3/4 vertebral fracture occurred during the correction, fortunately, there were no obvious perioperative complications, except for retropharyngeal swelling that required tracheotomy for a short time. We did not pay attention on vertebral artery injury immediately after the correction surgery, because the patient seemed to be normal. However, vertebral artery injury has been reported in 19% to 39% of all cases of cervical spine fracture.<sup>17,18</sup> In another study, multivariate regression analysis revealed that AS was a significant independent risk factor associated with vertebral artery injury, with an odds ratio of 8.04.<sup>19</sup> We underscore the importance of evaluation for vertebral artery injury immediately after the occurrence of vertebral fracture.

## CONCLUSION

We reported a rare surgical case of chin on chest deformity caused by upper cervical kyphosis associated with AS. Although sufficient correction was obtained with an unintentional vertebral fracture during surgery, the surgeon should be aware of iatrogenic fractures in a vulnerable spine of AS, because it may cause devastating neurological and vascular complications.

## CONFLICT OF INTEREST

The authors have nothing to disclose.

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