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Case Report

Chronic atlantoaxial rotatory fixation with neurofibromatosis type I: A case report

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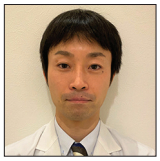

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ABSTRACT

Background: Atlantoaxial rotatory fixation (AARF) can be caused by infection, rheumatoid arthritis, surgery of head and neck, and congenital diseases. Type 1 neurofibromatosis (NF-1) is often associated with various musculoskeletal diseases, but few reports have described AARF with NF-1. Here, we report the success of a closed reduction and halo fixation utilized to treat chronic AARF with NF-1 in a 7-year-old female.

Case Description: A 7-year-old female with NF-1 presented with a 2-month history of torticollis and neck pain. C2 facet deformity had previously been identified on computed tomography (CT) before the onset of neck pain. Cervical radiography and CT showed AARF classified Fielding's Type I and Ishii's Grade II. Following 2 weeks of cervical traction, a closed reduction was followed by halo fixation that was utilized for 2 months. The patient fully recovered cervical range of motion following halo vest removal 4 months later. Further, the follow-up CT documented a normal atlantoaxial joint despite residual C2 facet deformity. In addition, no recurrence was evident 2 years later.

Conclusion: Halo fixation for chronic AARF with NF-1 proved effective. C2 facet deformity associated with NF-1 might have contributed to the onset of AARF.

Keywords: Chronic atlantoaxial rotatory fixation, Halo fixation, Neurofibromatosis type I

INTRODUCTION

Atlantoaxial rotatory fixation (AARF) is defined as dislocation or subluxation of the atlantoaxial joint. It occurs predominantly in children.[4] The clinical presentation typically includes neck pain, reduced range of motion, and torticollis (i.e., "cock-robin" position).[8] Most acute cases of AARF can be treated nonsurgically (i.e., with cervical traction and a collar).[4] However, rare cases of chronic AARF, typically defined as rotatory subluxation lasting longer than 2–3 months, may require surgical treatment.[2,6] Type 1 neurofibromatosis (NF-1) is an autosomal-dominant inherited disorder and often associated with various musculoskeletal diseases. Although spinal deformity in NF-1 is a common manifestation, cervical spine deformities requiring surgery are rare.[1,3,7] Here, we report the success of treating chronic AARF in a child with NF-1 utilizing closed reduction with protracted halo fixation.

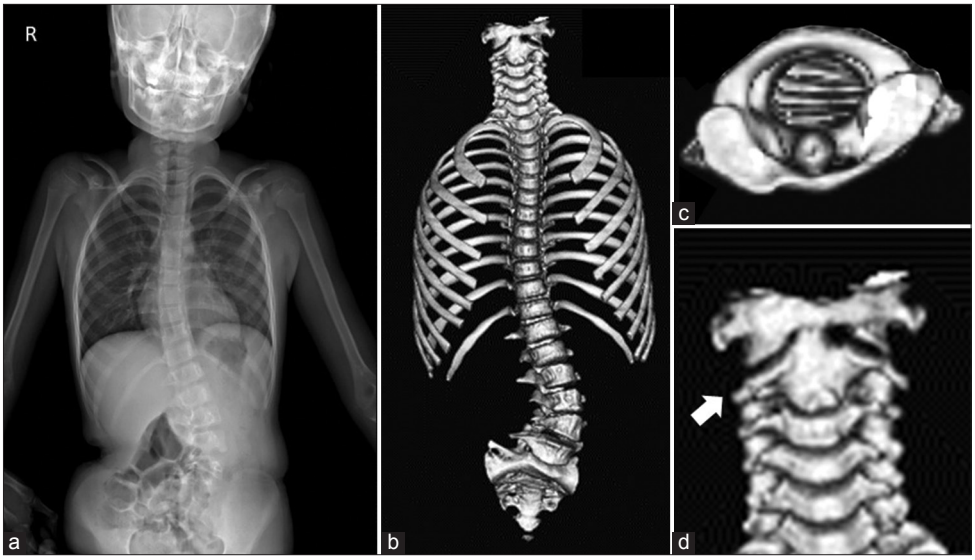

Figure 1: Before the onset of neck pain, whole spine radiography (a) and anterior view three-dimensional computed tomography (CT) (b) show the left-sided convex scoliosis curve. Cervical spine three-dimensional CT in the axial view (c) and anterior view (d) shows C2 facet deformity (arrow).

CASE PRESENTATION

A 7-year-old female with NF-1 presented with a 2-month history of torticollis and neck pain. She had been diagnosed with NF-1 due to the presence of scoliosis and café-au-lait spots. Radiographs previously showed a left convex scoliosis with a Cobb angle of 40º between L2 and L5 levels [Figure 1a]. A prior computed tomography (CT) had also demonstrated dystrophic changes, scoliosis, and C2 facet deformity [Figures 1b-d].

On examination, she had a left head tilt (i.e., the typical "cock-robin" posture). Cervical radiography and CT revealed that the C1-C2 joint was locked with C1 rotated 32° to the left and a C2 facet deformity [Figures 2 and 3]. These findings established the diagnosis of Fielding's classification type I and Ishii's classification Grade II AARF.[4,5] Magnetic resonance imaging showed no neoplastic lesions around the cervical spine.

Closed reduction and halo fixation

Three months after symptoms onset, a closed reduction and halo fixation were performed under general anesthesia [Figure 4]. Two months later, despite the CT showing a persistent C2 facet deformity, her cervical posture returned to the neutral position (i.e., only 4° of atlantoaxial rotation) [Figure 5], and the halo vest was removed. The follow-up CT showed normal alignment of the atlantoaxial joint, with 4° of the rotation, and C2 facet deformity 4 months after removal of the halo vest [Figure 6]. No further recurrence was identified 2 years later.

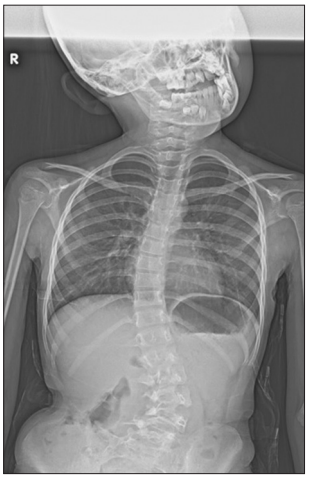

Figure 2: After the onset of neck pain, whole spine radiography shows lateral tilting of the cervical spine.

DISCUSSION

Causes of AARF include trauma, infection, rheumatoid arthritis, surgery of the head and neck, and congenital diseases (i.e., Down syndrome, Morquio syndrome, and Marfan syndrome).[6] Ishii *et al*. recommend careful closed manipulation followed by halo fixation for chronic AARF without C1-C2 bony union; the halo vest should be removed at a maximum of three postoperative months.[6] In addition, they found that C2 facet deformity was frequently observed in patients with chronic AARF, and remodeling of the C2 facet deformity helped to determine the appropriate duration of halo fixation.[6] In this case, no remodeling of the C2 facet deformity was found on the follow-up CT studies.

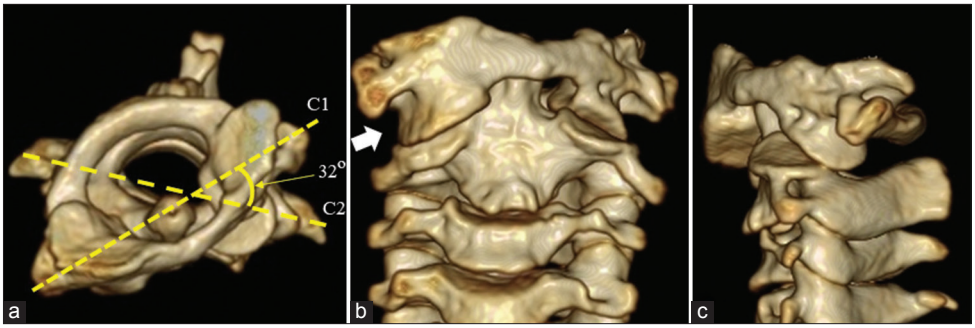

Figure 3: After the onset of neck pain, three-dimensional computed tomography in the axial (a), anterior (b), and lateral (c) views reveals that the C1-C2 joint is locked with C1 rotated 32° to the left and C2 facet deformity (arrow).

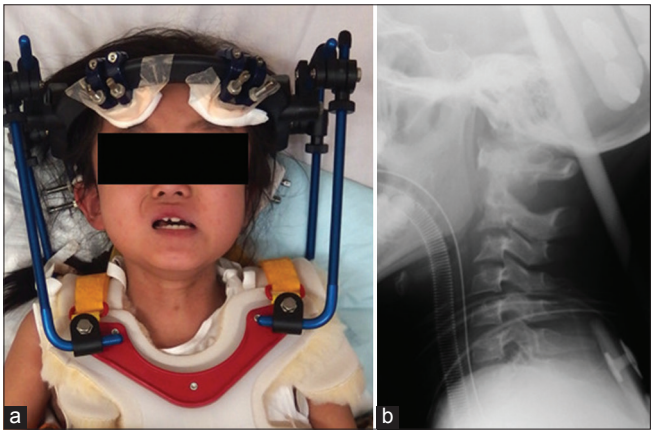

Figure 4: Photograph (a) and radiography (b) of the patient undergoing reduction treatment under general anesthesia.

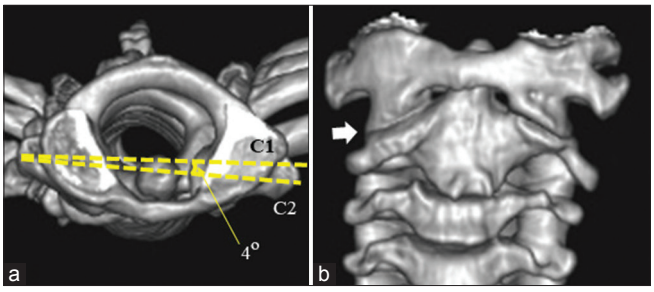

Figure 5: Two months after closed reduction and halo fixation, three-dimensional computed tomography in the axial (a) and anterior (b) views shows persistent C2 facet deformity (arrow), but cervical posture appears to have returned to a neutral position.

CONCLUSION

We successfully performed a closed reduction and halo fixation to treat chronic AARF in a child with NF-1.

Ethical approval

Informed consent was obtained from the patient and her family before data use, and patient confidentiality was assured.

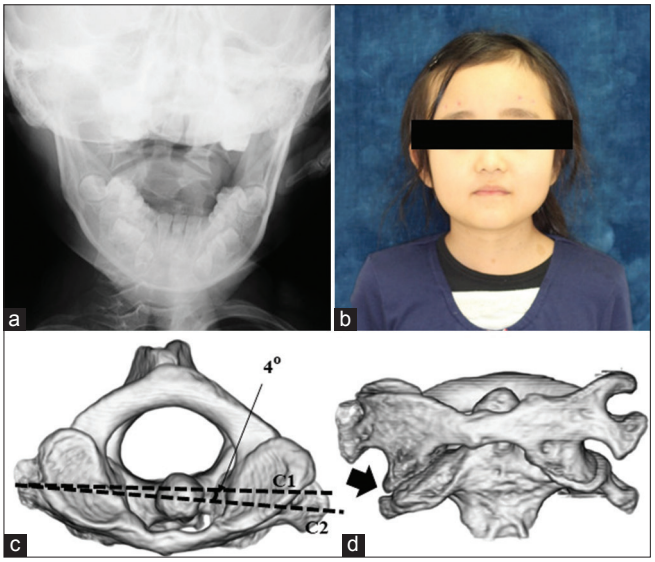

Figure 6: Photograph (a) and radiography (b) at 1 month after removing halo fixation. Three-dimensional computed tomography images in the axial (c) and anterior (d) views at 4 months after removing the halo fixation show normal alignment of the atlantoaxial joint and C2 facet deformity (arrow).

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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