Case Report

Navigation-assisted occipitocervical fixation and decompression in a patient with polyostotic fibrous dysplasia

ABSTRACT

Fibrous dysplasia (FD) is a rare skeletal disorder characterized by the replacement of normal bone with fibrous connective tissue, leading to abnormal bone formation. This case report details the successful treatment of a 61-year-old woman with FD at the craniovertebral junction (CVJ). The patient, who had a history of intracranial meningioma and had already been diagnosed with FD, experienced worsening gait disturbance and muscle weakness following a fall. Imaging studies revealed extensive polyostotic FD lesions in the skull and cervical spine, along with a C2 odontoid fracture causing spinal cord compression. The patient underwent occipitocervical fixation and decompression surgery. Intraoperative O-arm navigation was used to ensure accurate screw placement and effective decompression. This procedure allowed for proper positioning of the C2 and C3 pedicle screws, resection of the hyperplastic occipital bone and C1 posterior arch, and placement of the occipital plate with avoiding the cyst components. At a 2-year follow-up, there were no signs of screw loosening, and the patient showed marked clinical improvement. This case emphasizes the importance of tailored surgical strategies and the use of advanced navigational technologies in managing complex FD cases, particularly those involving the CVJ. It also highlights the challenges of treating polyostotic FD, where complete resection is often unfeasible. The successful outcome in this case supports the use of decompressive surgery combined with stabilization to relieve symptoms and prevent further complications.

Keywords: Craniovertebral junction, fibrous dysplasia, upper cervical

INTRODUCTION

Fibrous dysplasia (FD) is a skeletal disorder characterized by the replacement of normal bone with fibrous connective tissue, resulting in abnormal bone formation. FD can affect any bone in the body.[1] When FD involves the craniovertebral junction (CVJ), it can cause neurological symptoms resulting from compression of the spinal cord or brainstem. Surgical decompression and stabilization may be necessary to alleviate symptoms and prevent further complications. Here, we present a rare case of occipitocervical fixation and decompression in a patient with polyostotic FD with a C2 odontoid fracture leading to myelopathy.

CASE REPORT

A 61-year-old woman who had already been diagnosed with FD was being followed up at another hospital for an intracranial meningioma. She was referred to our hospital 3 months after falling from a standing height and worsening gait disturbance. Physical examination revealed mild muscle weakness in the left upper extremity. Deep tendon reflexes of the upper and lower extremities were hypertonic, and Hoffman's sign was positive on the left side.

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Submitted: 02-Jul-24 **Accepted:** 31-Jul-24
Published: 12-Sep-24

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.jcvjs_104_24	

How to cite this article: Nagashima Y, Nishimura Y, Abe T, Saito R. Navigation-assisted occipitocervical fixation and decompression in a patient with polyostotic fibrous dysplasia. J Craniovert Jun Spine 2024;15:380-3.

Radiographs showed extensive polyostotic FD lesions in the skull and cervical spine [Figure 1]. Furthermore, computed tomography (CT) images showed a C2 odontoid fracture, and there is a bony fusion between the skull and C1 and the bone fragments of C2, as well as between C2 and C3. Posterior views of the three-dimensional CT image revealed the posterior arch of C1 was not visible due to the expanded occipital bone. The sagittal T2-weighted magnetic resonance imaging (MRI) image (D) revealed that the posterior arch of C1 is compressing the spinal cord, causing high signal changes on the dorsal aspect of the spinal cord [Figure 2].

The patient underwent occipitocervical fusion and decompression. Intraoperative navigation with the O-arm system was employed for accurate screw insertion. First, C2 and C3 pedicle screws were inserted. Then, decompression of the foramen magnum was performed. Due to the hyperplasia of the occipital bone, C1 was initially not visible, so navigation-guided resection of part of the occipital bone, followed by resection of the posterior arch of C1 [Figure 3]. Then, the occipital plating was performed. The occipital bone was trimmed with a drill, and screw positions for the occipital bone plate were determined to avoid the cystic portion of the occipital bone [Figure 4]. The bone graft placement was not performed.

Postoperatively, cervical X-ray and CT image revealed the implant was placed in the correct position, and the MRI image confirmed adequate spinal cord decompression [Figure 5]. Subsequently, the patient underwent an intracranial meningioma resection. At a 2-year follow-up, there were no signs of screw loosening, and the patient was clinically markedly improved.

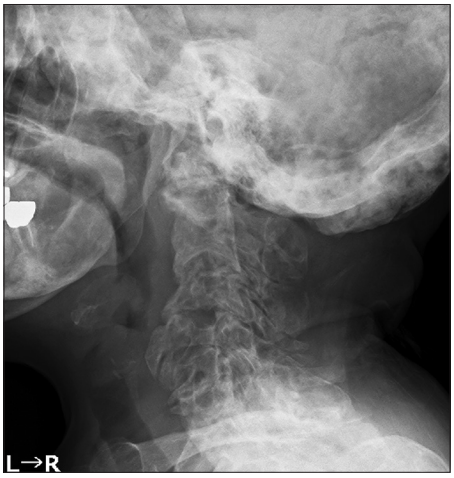

Figure 1: The lateral view of the cervical spine shows vertebral deformity and cystic degeneration of the vertebral bodies and spinous processes. The sphenoid and occipital bones are affected and grossly enlarged

DISCUSSION

FD is a bone disorder in which immature, haphazardly distributed fibro-osseous tissue replaces normal bone, affecting the skull, facial bones, femur, tibia, ribs, humerus, and pelvis.[1] FD exhibits a mosaic pattern and manifests across a wide clinical spectrum, affecting either one bone (monostotic) or multiple bones (polyostotic).[2] FD may be associated with endocrine disorders and skin lesions as part of McCune–Albright syndrome. Approximately 60% of patients with polyostotic FD have symptoms by 10 years of age.[3] FD often presents with pathological fractures.

FD occurs rarely in the cervical spine in approximately 2.5% of FD cases.[4] A case review report collecting FD of the cervical spine reported 22 cases (62%) of monostotic FD and 14 cases (38%) of polyostotic FD, with only 10 cases presenting with neurologic symptoms.[5] Some authors

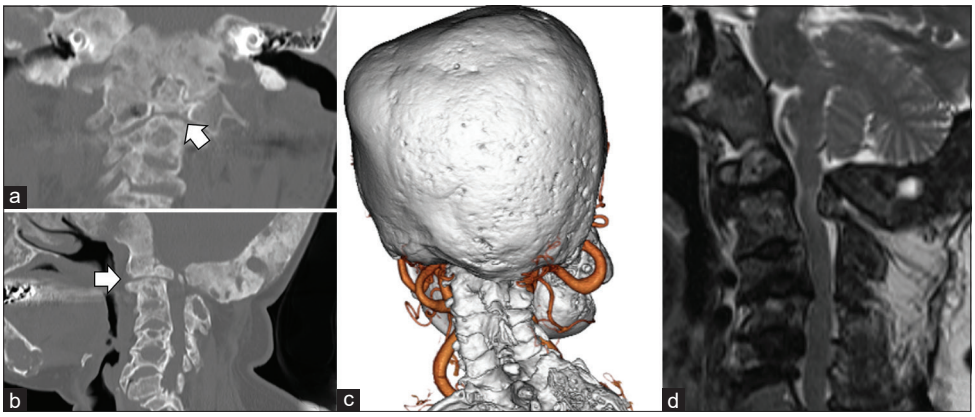

Figure 2: Coronal (a) and sagittal (b) computed tomography images show a mixture of sclerotic and cystic lesions in the skull and cervical spine. In addition, there is a fracture of the C2 odontoid process (arrow). The skull bone, C1, and C2 fragments are fused, as are C2 and C3. In the three-dimensional CT image, the posterior arch of C1 is not visible from the dorsal view due to the proliferated occipital bone (c). The sagittal T2-weighted magnetic resonance imaging image (d) shows the posterior arch of C1 compressing the spinal cord, resulting in high signal changes in the spinal cord

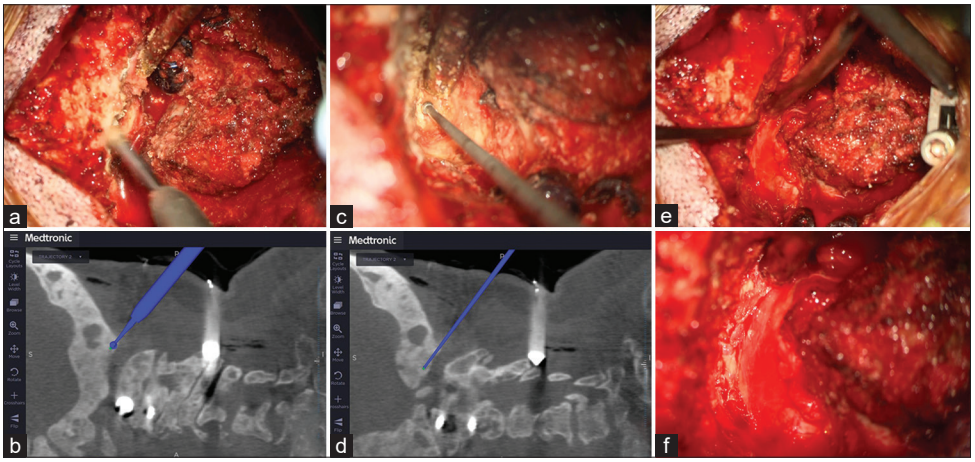

Figure 3: Intraoperative images of the surgical procedure and O-arm navigation during C1 posterior arch resection. Drilling of the occipital bone was performed under navigation guidance (a and b), with periodic confirmation of the position using a navigation probe (c and d). After exposure of the C1 posterior arch, bone resection and neural decompression were achieved (e and f)

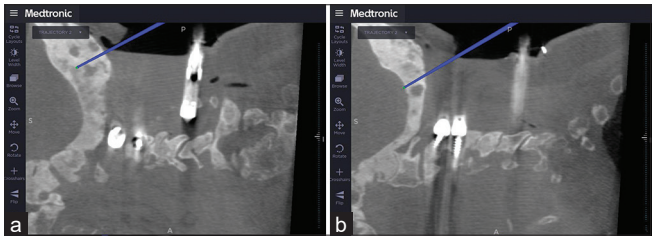

Figure 4: The navigation probe was inserted into the pilot hole of the screw for placement of the occipital bone plate to ensure that the cyst areas were avoided (a and b)

advocate for radical resection of the affected area to prevent recurrence and malignant transformation.[6] This strategy may be effective when the lesion is localized. However, radical resection is not feasible for some patients, particularly those with polyostotic FD.[6] Therefore, treatment options, including decompression surgery, vertebral body reconstruction, and stabilization with or without complete resection, are determined on a case-by-case basis. In our case, lesions extended from the skull to the cervical spine and throughout the body, making complete resection impossible. Since FD is basically a benign tumor, surgical intervention is aimed at relieving persistent pain, neurologic deficits, vertebral collapse, and instability.

Cervical FD can sometimes be revealed through pathological fractures. Among the cases of cervical FD identified through pathological fractures, C2 fractures were the most frequently reported.[7] Treatment approaches for FD of the C2 fractures include posterior fixation, vertebroplasty, and resection of the lesion, followed by bone grafting. However, there is a scarcity of reports regarding therapeutic interventions for extensive lesions involving both the skull and the cervical spine, as observed in our case.

In our case, we opted for posterior craniocervical fixation and decompression. Although we faced some challenges due to anatomical abnormalities, the use of O-arm navigation for drilling and placement of the occipital plate, as reported in previous studies,[8-10] enabled us to safely complete the treatment. It was necessary to resect a portion of the proliferated occipital bone during the C1 posterior arch resection. In addition, the placement of the screws presented difficulties. FD manifests in various forms depending on the location, including ground-glass, sclerotic, and soft-tissue components. Utilizing O-arm navigation, we were able to avoid soft-tissue components and accurately place the occipital bone plate. Thus, O-arm navigation proved the resection of the lesion and the placement of the occipital bone plate and pedicle screws.

CONCLUSION

We successfully treated a patient with FD at the craniovertebral junction using occipitocervical fixation and decompression surgery. Postoperative results showed significant improvement and stable fixation over 2 years. O-arm navigation facilitated accurate screw placement and effective decompression.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Acknowledgment

This manuscript underwent English language corrections

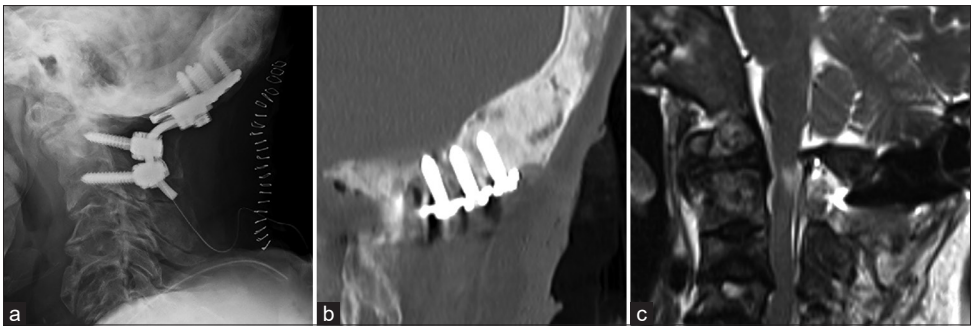

Figure 5: Postoperative lateral X-ray showed that the implant was placed in the correct position (a). The computed tomography lateral view showed that the occipital bone screws were inserted with sufficient length (b) and the magnetic resonance imaging T2 sagittal image showed adequate decompression of the spinal cord (c)

using ChatGPT (GPT-4o version). All data interpretation and final manuscript revisions were conducted by human researchers.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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