Eur Spine J
DOI 10.1007/s00586-017-5083-x

CASE REPORT

Traumatic multiple cervical spine injuries in a patient with osteopetrosis and its management

Ashok Keshav Rathod[1] · Rakesh Padmakar Dhake[1] · Mandar Deepak Borde[2]

Received: 20 October 2016 / Accepted: 1 April 2017

Abstract
Study design Single case report.
Objective/purpose To report multiple level fractures of cervical spine in a patient with osteopetrosis and its management.
Summary of background data Osteopetrosis is a rare inherited condition characterized by defective remodeling resulting in hard and brittle bones with diffuse osteosclerosis. Fractures of spine are rare as compared to the common long bone fractures. We report a case of traumatic multiple level fractures of cervical spine in osteopetrosis and its management which has rarely been reported in the literature before, if any.
Methods 17-year-old boy presented with severe tenderness in neck and restricted range of motion following a trivial injury to the neck in swimming pool. The neurology was normal and he was diagnosed to have autosomal dominant osteopetrosis on evaluation. Imagining findings, clinical course and the method of treatment are discussed.
Results Radiological evaluation revealed presence of multiple level fractures of cervical vertebrae with end plate sclerosis. Patient was managed with cervical skeletal traction in appropriate extension position for 6 weeks followed by hard cervical collar for another 6 weeks. Follow-up radiographs at 18 months and 2.5 years showed healed fractures with no residual instability or symptoms.
Conclusion The case report discusses rare occurrence of multiple level fractures of cervical spine following trivial injury to the neck in a patient with osteopetrosis and its treatment with conservative management.

Keywords Osteopetrosis · Multiple fractures · Cervical spine · Conservative management

Introduction

Osteopetrosis is a rare condition characterized by marked radiodensity of bones throughout the skeleton [1] and poor remodeling due to dysfunction of osteoclast mediated bone resorption. As with many hereditary skeletal disorders it is not a single disease but a syndrome with several variants [1]. Broadly, it is categorized into an infantile malignant autosomal recessive form and an adult benign autosomal dominant form. The infantile form worsens rapidly and in absence of effective medical intervention, leads to death in the first few years of life [1]. The autosomal dominant form is usually asymptomatic and many patients are not diagnosed until the appearance of bone fracture [2]. Here we discuss the rare occurrence of multiple traumatic cervical vertebral fractures in a patient diagnosed to have osteopetrosis following the injury and its management. After an extended search, we found no similar case of traumatic multi-level fractures of cervical vertebrae in osteopetrosis being reported in the English language

✉ Rakesh Padmakar Dhake
drrakeshdhake@gmail.com

Ashok Keshav Rathod
akrkem@gmail.com

Mandar Deepak Borde
mandarborde@gmail.com

[1] Department of Orthopaedics, LTMMC & LTMGH, First floor, College building, Sion hospital, Sion west, Mumbai, Maharashtra 400022, India

[2] Spine Fellow, Lilavati Hospital and Research Centre, Bandra reclamation, Bandra west, Mumbai, Maharashtra 400050, India

Published online: 09 April 2017

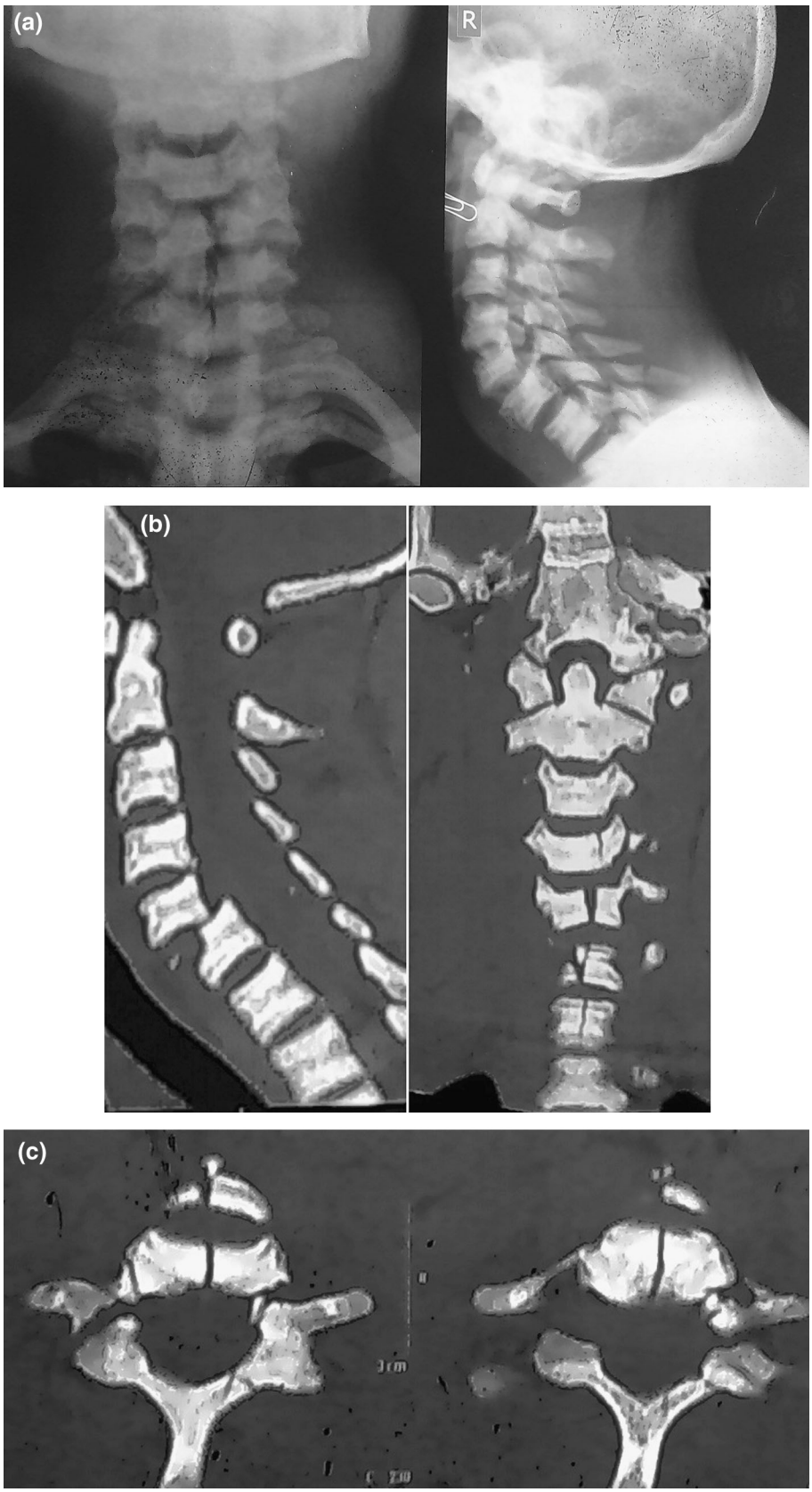

Fig. 1 **a** Plain X-ray and **b**, **c** CT scan showing multiple cervical vertebral fractures

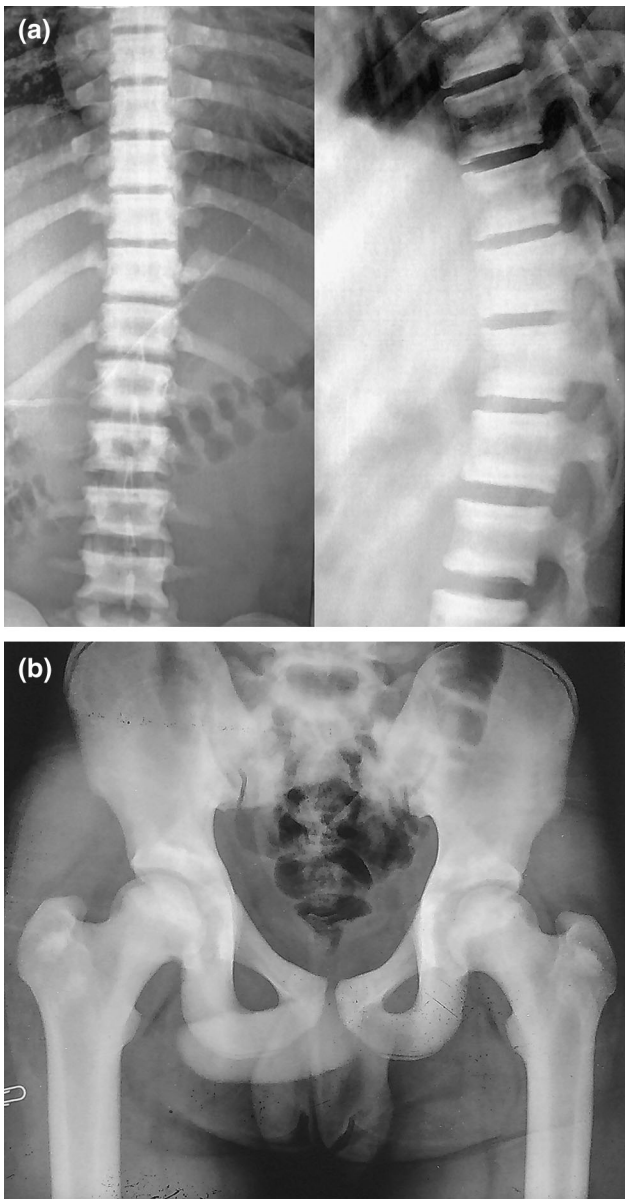

Fig. 2 Diffuse involvement of axial skeleton **a** "sandwich" vertebrae, **b** pelvis

Fig. 3 Symmetrical involvement of long bones

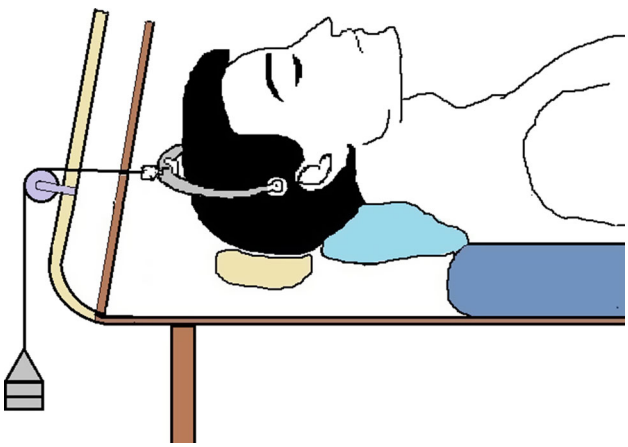

Fig. 4 Illustration showing application of cervical traction

literature. It was, therefore, considered worth reporting such complex cervical injury in an osteopetrotic patient, its treatment and outcome. The aim of describing this case scenario and technical tips is to help readers faced with similar problems.

Case report

A 17-year-old boy was presented to the emergency department following a trivial injury. He was swimming in a pool when another person jumped into the water, and accidently landed his both knees on his neck. Sudden severe pain in the neck led to discontinuation of swimming and he was taken to hospital for the same.

On examination, the patient had no associated injuries elsewhere and was hemodynamically stable. Tenderness in the neck was associated with restricted range of motion and no neurological deficit. A hard cervical collar was applied and patient was subjected to radiological evaluation. The Computerized tomography (CT) scan showed presence of multiple fractures of the cervical vertebrae with vertical split of C4–C7 bodies, fracture of posterior elements of

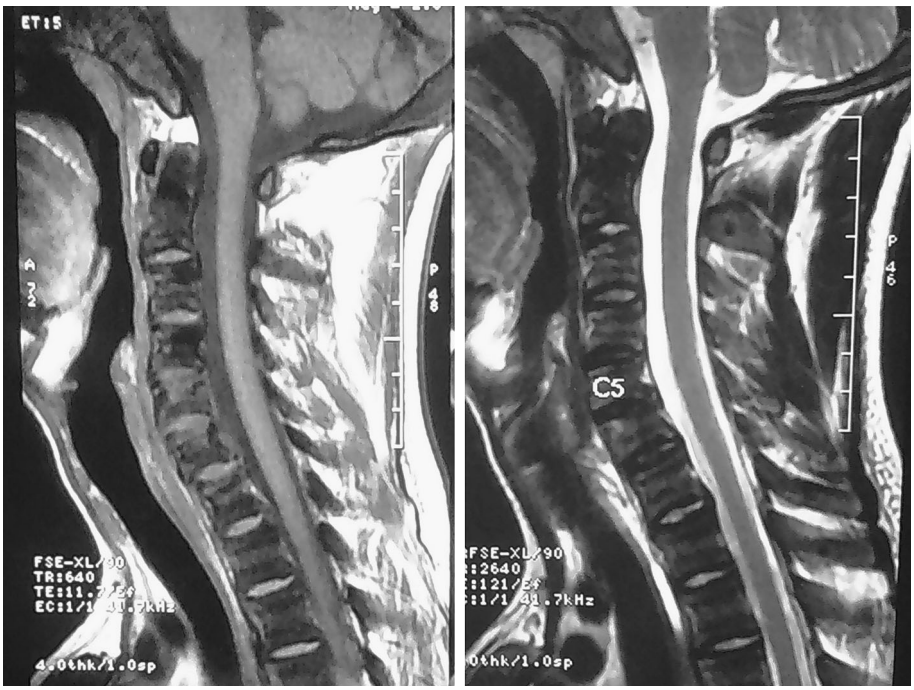

Fig. 5 MRI at 12 weeks after injury

C3–C7 and C5–C6 unifacetal dislocation with anterolisthesis (Fig. 1).

As the X-rays showed marked sclerosis of the vertebrae, patient was subjected to further radiological and hematological investigations to rule out other causes for osteosclerosis. Patient did not have any hematological abnormalities and the blood biochemical values were within normal limits except the elevated acid phosphatase levels (15 U/L). X-rays showed diffuse osteosclerosis primarily involving the axial skeleton with symmetrical involvement of long bones (Figs. 2, 3). Vertebral endplate sclerosis was seen with well-demarcated margins between the peripheral sclerotic and central relatively lucent bone, consistent with a "Sandwich vertebrae" appearance. The visualized lumbar vertebrae also demonstrated a "bone within a bone" appearance secondary to sclerosis along the inner margins of the lateral aspect of the vertebral bodies, along with the endplate sclerosis. All these radiographic findings along with the elevated acid phosphatase levels were consistent with autosomal dominant osteopetrosis [1, 3, 4].

The injury extended from C2 to C7 vertebral level. It was expected that surgical stabilization involving one normal level above and below the injured segments would amount to fixation of the entire cervical spine (probably leading to an occipito—D1 construct), thus compromising its mobility to a large extent. Also, this long segment fixation would be accompanied by the added complications of surgical intervention in an osteopetrotic spine. We thus aimed at achieving the stability by allowing the cervical spine to fall into reasonably acceptable alignment with close reduction of the facetal dislocation and maintaining the alignment with traction as the soft tissue healed. Reassessment of the stability would then be considered and any residual subluxation be dealt with fixation of that particular segment, thus avoiding fixation of an entire cervical spine.

Traction was achieved with weights applied to Gardner–Wells tongs and suspended freely over a pulley attached to head end of the bed (Fig. 4). Initial weight of 10 lbs was applied in a flexed position. Increments of 5 lbs were made at an interval of 20 min followed by a lateral shoot through X-ray and neurological assessment. After reduction was achieved, neck was placed in extension with lighter maintenance weights. Mattress was pulled towards the feet, ending at shoulder level and the head was supported with a soft cushion or a water filled glove under the occiput preventing any pressure sores. Head rested at a lower level than shoulders, thus maintaining the neck in an extended position. Counter traction was achieved by elevating the head end of the patient. Adequate skin care at the pin insertion sites was ensured.

Continuous traction in addition to causing reduction of facetal dislocation also produced a relative fixation of injured segments by the rigidity conferred by surrounding soft tissue structures under tension. Reduction of facetal dislocation allowed the cervical spine to fall into fairly acceptable alignment which was supervised with weekly lateral X-rays.

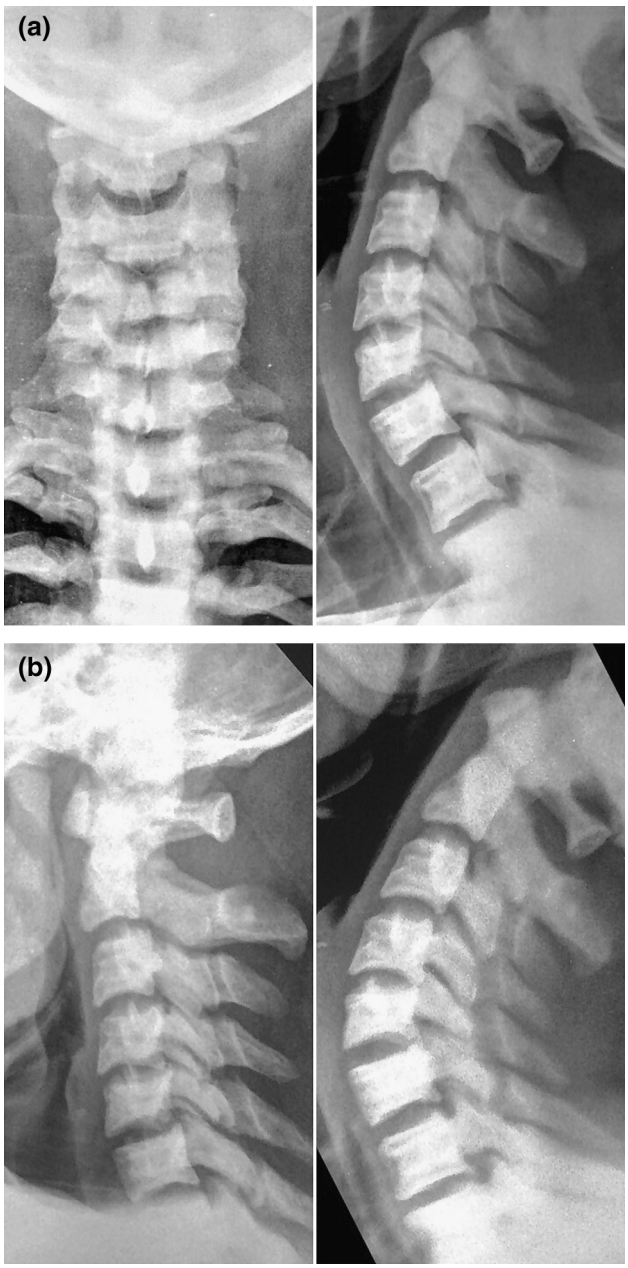

Fig. 6 Follow-up X-rays at 18 months after injury **a** antero-posterior and lateral, **b** flexion and extension views

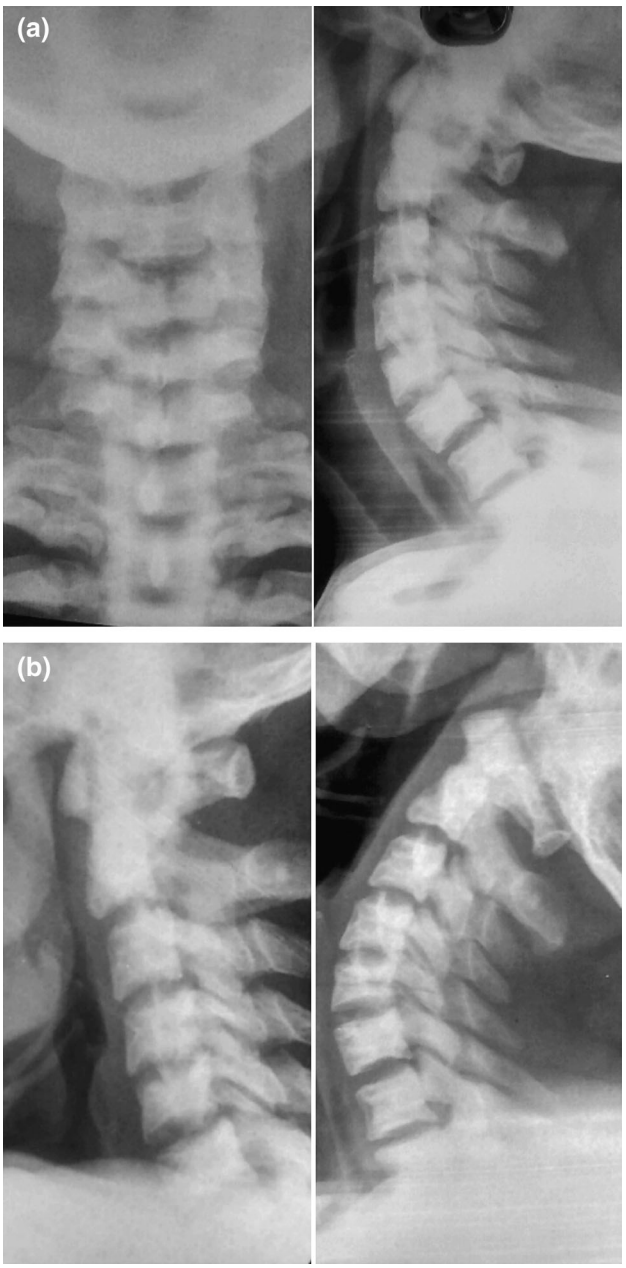

Fig. 7 Follow-up X-rays at 2.5 years after injury **a** antero-posterior and lateral, **b** flexion and extension views

With healing of the soft tissues after 6 weeks, tongs were removed and the stability was reassessed. A flexion–extension radiograph obtained, showed no residual instability which would otherwise require surgical intervention as anticipated. Encouraged with these early good results, traction was discontinued and the patient was permitted to ambulate with an extended period of immobilization to another 6 weeks in a hard cervical collar. A repeat radiograph showed well consolidated fracture sites and the cervical collar was thus discontinued. MRI was then performed to further evaluate the injury (Fig. 5). The vertebral bodies and the intervertebral discs were fairly normal in height and intensity. No foraminal narrowing or spinal cord compression was observed.

Follow-up at 18 months and 2.5 years with flexion–extension radiographs suggested no residual instability, and the patient was completely asymptomatic, with preserved range of motion (Figs. 6, 7, 8). Repeat MRI at 2.5 years showed no residual facetal subluxation or any obvious reduction in the injured C5–C6 disc space, contrary to the natural history of an injured disc (Fig. 9). Capsular and soft tissue healing over a period of time led to an apparent

Fig. 8 Range of motion at follow up **a** flexion and extension, **b** lateral bending, **c** rotations

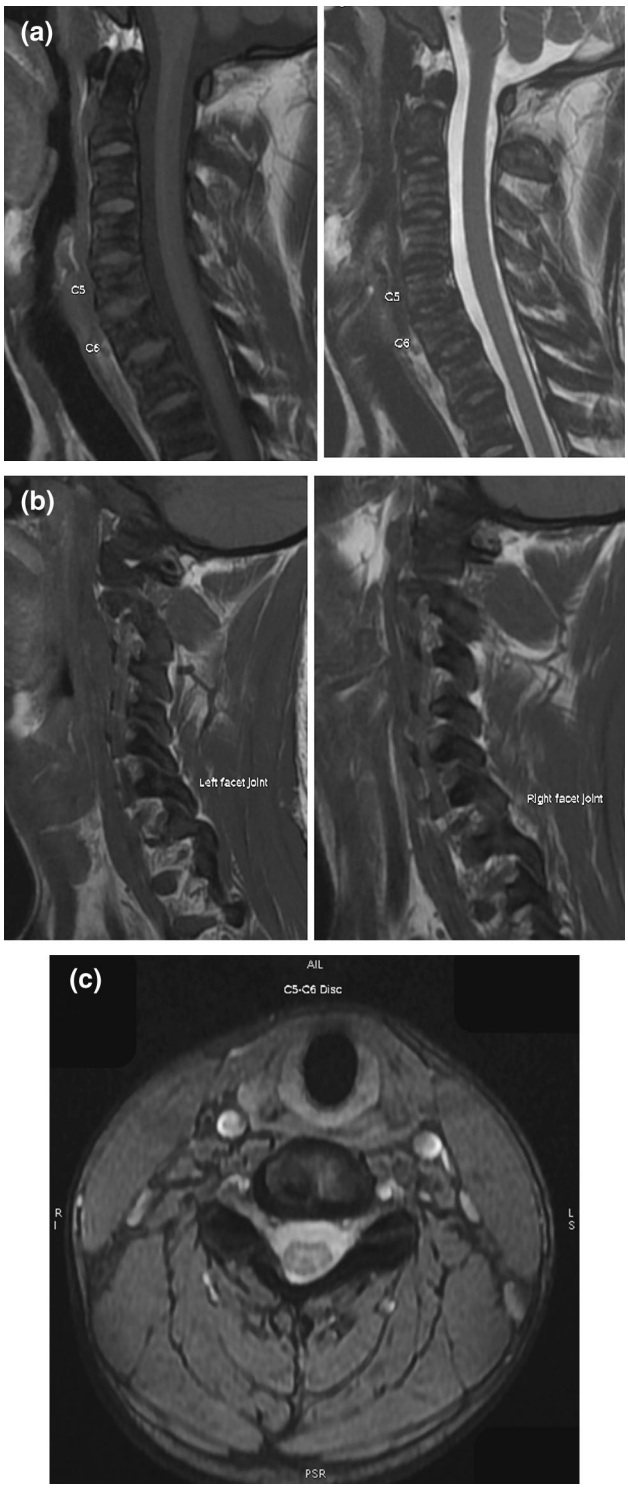

Fig. 9 Follow-up MRI at 2.5 years after injury **a** T1 and T2 sagittal views, **b** left and right facet joints, **c** axial view at the level of C5–C6 intervertebral disc

restabilization of the injured C5–C6 segment, and we presume that in long run, this disc will probably degenerate.

Discussion

We discuss the rare occurrence of traumatic multiple level fractures of cervical spine following trivial injury to the neck in a patient with osteopetrosis. Fracture is a common complication in osteopetrosis but spiral, compression or comminuted fracture is rare [2]. It seems that the osteopetrotic bone resists the force in parallel with the cortical layer, but is fragile to forces at right angles to the direction of cortical layer. Most fractures are thus of transverse types and the fracture line is perpendicular to the direction of cortical layer [2]. Our patient had a vertical split fracture of the C4–C7 vertebral bodies at right angles to the sclerosed endplates. The posterior elements fracture line also was at right angles to the cortical layer.

Historically most of the fractures in osteopetrosis have been treated conservatively [5]. Because the healing response is variable, management must be adapted and individualized to address the technical challenges unique to this patient population. Operative intervention has a high rate of intraoperative and post-operative complications [6]. Cervical fractures and instability in patients with osteopetrosis represent a unique challenge to the orthopedic surgeons. Thus, a good planning and anticipation of possible technical difficulties and associated complications is mandatory in such patients and a decision should be taken after weighing the risk benefit ratio. De Palma et al. [7] in a report on histology of fracture callus in a patient with autosomal dominant osteopetrosis found that, at 1 year of healed fracture, biopsy showed unorganized woven bone with absence of osteoclasts. Thus, osteopetrotic fractures do heal but remodeling does not occur. The fact that osteopetrotic bone heals despite defective remodeling and Haversian system makes the contribution of periosteum in fracture healing a potential area of research in such patients.

We thus report a case in which intervertebral cervical discs quite obviously survived a severe discoligamentous injury—as documented by the MRIs. In times where surgical aggressiveness is at a high and where such injuries are typically recommended for interbody fusion at primary surgery, we believe that this case would be a stimulus for a much necessary discussion. Granted, the case is on a young adult with a condition that has altered bone biology, but nevertheless it demonstrates in a beautiful way what conservative care together with surgical patience and restraint can accomplish. Also, it is no small difference for this patient to remain with a mobile cervical spine as opposed to be entering adult life with a multilevel fusion.

Conclusion

In our patient having multiple fractures of cervical vertebrae following a trivial injury to neck, complete recovery was seen after conservative treatment with cervical traction followed by hard cervical collar, with no residual instability at 2.5 years of follow up. However, the possibility of any deformity or intervertebral disc degeneration is to be looked for during long-term follow-up, and education of the patient is necessary to prevent further spinal injuries that may be brought about due to fragile bones.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

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