C2/C3 pathologic fractures from polyostotic fibrous dysplasia of the cervical spine treated with percutaneous vertebroplasty

David Dang · Mirza N. Baig · Greg Christoforidis ·
E. Antonio Chiocca · Joshue Gabriel

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Abstract We will discuss a potential role of percutaneous vertebroplasty (PVP) in the management of patients with severe fibrous dysplasia of the spine with multiple cervical lesions and C2–C3 pathologic fractures that may not be a good surgical candidate. Polyostotic fibrous dysplasia involvement of the cervical spine is rare. Review of literature indicates only few reported cases of surgical management with one case of mortality indicating increased risks associated with surgical intervention. While PVP is commonly used for the treatment of osteoporotic thoracolumbar vertebral compression fractures, its role in vertebral stabilization for fibrous dysplasia has not been reported. A 35-year-old man with McCune–Albright syndrome and severe polyostotic fibrous dysplasia of C2 and C3 vertebrae presented with severe neck pain, radiculopathy, quadriplegia and myelopathy. The lesion had pathologic fractures, and there was an os odontoideum with cervical cord atrophy at the C1 level. After discussing need for aggressive surgical management and potential complications, we offered PVP due to surgical risks involved. PVP was performed with a posterolateral transpedicular approach without complication. The patient had remarkable improvement in clinical relief of neck pain and

improvement of myelopathic symptoms at 1-year follow-up. We present a case that illustrates a potential use of PVP in the management of a patient with symptomatic spinal fibrous dysplasia with associated pathologic fractures who was poor surgical candidate.

Keywords Percutaneous vertebroplasty · Polyostotic · Fibrous dysplasia

Introduction

Fibrous dysplasia is relatively uncommon fibro-osseous abnormality of bone with unknown etiology. In certain individual with genetics predisposition multiple bones can be affected especially in syndromes including McCune–Albright. Vertebral bone involvement is seen in small percent (~4%) of polyostotic fibrous dysplasia [7]. In patients with polyostotic fibrous dysplasia, involvement of the cervical spine is uncommon with less than 20 reported cases. In the majority of the patients the lesion is incidentally discovered [5]. Surgery is performed when the patient has persistent pain, neurologic impairment, vertebral collapse, instability, and/or cord compression [8]. Surgery however is complicated as the involved bone is brittle. Although a consensus for management of this disease has not been achieved, many authors recommend radical removal of all involved bone as well as internal fixation or bone graft-assisted fusion to achieve long-term stabilization. While this is the goal, it may not be feasible in all cases, and case-by-case management is recommended. We present the management and clinical course of a poor surgical candidate with fibrous dysplasia lesions and pathologic fractures of C2–C3 complex using percutaneous vertebroplasty (PVP).

M. N. Baig (✉) · E. A. Chiocca
Neurological Surgery, Ohio State University,
410 West 10th Street, N1014 Doan Hall, Columbus,
OH 43210, USA
e-mail: doctorbaig@gmail.com

J. Gabriel
Orthopedics Department, Ohio State University,
Columbus, OH, USA

D. Dang · G. Christoforidis
Department of Radiology, Ohio State University,
Columbus, OH, USA

Case report

A 35-year-old man with McCune–Albright syndrome with severe polyostotic fibrous dysplasia was referred to our institution for the management of neck pain, paresthesias, and progressive quadriparesis. He had extensive craniofacial deformity and experienced occasional pre-syncope with cervical motion. His symptoms had progressed over a period of 1 year, and at presentation he had to manually support his head to avoid neck pain. The patient was dependent for all daily living activities (any time of disability).

Neurological examination demonstrated limited range of cervical motion and positive Lhermitte's sign with pain and lightheadedness. Spurling's and Hoffman's signs were positive on both sides. There was decreased pinprick and vibratory sensations in all extremities. Extremity strength exam indicated good strength in all groups with possible slight unexplained weakness (five out of five) on right upper extremity. Reflex exam indicated symmetrical 3+ hyperreflexia throughout.

Computed tomography (CT) (Fig. 1) demonstrated C2–C3 fusion with a lytic expansile lesion. There were few small pathologic fractures of the vertebral body complex in both anterior and inferior bone cortices. Aside from the

large lytic expansile lesion there was also findings of an os odontoideum. It was difficult to evaluate for cervical instability as the patient could not adequately perform flexion and extension radiographs. From the CT images, it was felt that the os odontoideum was fixed to anterior arch of C1.

MR of the cervical spine (Fig. 2) indicated an expansile lytic lesion isointense on MR T1-weighted sequence and hyperintense on T2-weighted sequence. There was mild gadolinium enhancement. While there were no signs of signal changes on the spinal cord, it was significantly atrophic at the medullary–cervical junction.

The patient was felt to be at risk of a C1–C2 level quadriplegia as the massively enlarged skull and brittle vertebrae would lead to fracture and cord injury. Surgical options included a combined posterior occipitocervical fusion with C1–C2 laminectomy and anterior fusion from the clivus to C4 vertebrae. Surgical treatment and its potential complications were discussed with the patient. Due to the risks involved with such procedure, alternative treatment options were also investigated. We considered PVP as a potential management for mechanical support and stabilization of C2–C3 fibrous diseased bone with potential treatment of pathologic fractures within this segment thus providing pain control. The patient opted for PVP as an

Fig. 1 Sagittal and coronal CT images (a, b) and lateral radiograph (c) show a multiseptated C2–C3 vertebral lesion with disruption of the anterior and inferior cortices of the fused vertebral body (arrows). Axial postmyelogram CT (d) shows marked spinal cord atrophy at the C1 level. Note large dysplastic calvarial bones

Fig. 2 MR sagittal T1-weighted with gadolinium (a) and sagittal (b) and axial (c and d) T2-weighted images show typical appearance of a fibrous dysplasia lesion involving the C2–C3 complex, containing mainly of fluid-like components. There was some enhancement at the periphery. While there were no signs of signal changes in the spinal cord, it was significantly atrophic at the medullary–cervical junction

initial step with possible unavoidable need for surgical intervention in the future.

The procedure was performed under general anesthesia with somatosensory evoked potential monitoring. Under CT guidance a posterolateral transpedicular approach was performed with a trocar-cannula needle placed through the left C3 pedicle into the lesion with the patient placed in the supine position (Fig. 3). Once this was done, under biplane fluoroscopy, a catheter with its tip cut off was advanced into the lesion, using Seldinger's technique with the back end of a 0.035-in. guide-wire.

Polymethylmethacrylate (PMMA) material was injected via the catheter. Post-procedural CT showed a column of

Fig. 3 Intra-procedural CT demonstrates access to the C2/C3 vertebral complex via the left C3 pedicle

cement formed from the top to the bottom of the lesion (Fig. 4).

The patient's neck pain resolved one week after the procedure. His extremity strength and cervical range of motion improved, and by the fifth week he no longer required a neck brace. He continued to improve at the 6-month and 1-year visits; he was able to participate in all activities of daily living and yard work.

Discussion

Fibrous dysplasia is an uncommon benign fibro-osseous abnormality of the bone. Cervical spine involvement is rare [7]. Surgical management has been reported in only seven patients who had pathologic fracture, neurologic deficits, persistent neck pain with or without deformity [2, 4, 6, 8, 9, 12, 15]. Surgical approach is advocated to be tailored to the involvement of the dysplastic tissues. The disease is however often diffuse and combined posterior and anterior stabilization has been performed [6, 7, 9, 12]. Curettage with bone graft placement can be performed but the dysplastic tissues have been found to extend into the grafted materials. [5, 14] The risks involved with surgery of cervical spine fibrous dysplasia can be potentially high. The bone is brittle as the cortex may be eggshell thin [2, 5, 6, 15]. The anatomy may be so distorted by the dysplastic tissues that the interlamina spaces

Fig. 4 Post-procedural CT images show deposition of the bone cement in multiple compartments of the vertebral complex lesion and formation of a column of cement from top to bottom

can be widened, the ligamentum flava can be absent and the dura-arachnoid tissues can be denuded [12]. Mortality can occur, particularly when there is significant deformity [12].

We present a potential treatment of pathologic fractures with associated myelopathy from vertebral fibrous dysplasia with PVP. This technique has been used for the treatment of osteoporotic vertebral collapse, vertebral angiomas, and malignant tumors [3]. It provides mechanical support and pain control. The complication rate of PVP is low. We project that the complication rate for fibrous dysplasia treatment would be less than or comparable to that of angioma, which is approximately 2–5%. The main complication is radiculopathy from cement leakage into the epidural or foraminal veins [3].

Most investigators have expressed that the presence of symptoms from spinal cord compression is a contraindication to the performance of PVP [13]. This case report does not refute this belief. Other investigators had performed PVP successfully in patients with spinal canal compromise with vertebral metastasis [1, 11]. The patients selected in these studies however did not have spinal cord symptoms. We believe there is a potential for furthering the role of PVP in the management of symptomatic spinal disease, and there is a need for further experience.

While the mechanism of neurological improvement in our patient is unclear, it is possible that the cement reduced micro-motion of the fractured lesion and atlanto-axial

movement. Also, the PMMA might have resulted in cell death or reduced metabolic activity of the lesion and improved vascular flow to the spinal cord. This notion is supported by a study that showed that proliferation of new neoplastic cells was rare in the vertebrae already cemented [16]. One autopsy study even demonstrated cell death in a treated vertebral body [10].

Conclusions

In summary we present a potential role for PVP in the management of spinal disease. We treated a patient with C2–C3 pathologic fractures due to polyostotic fibrous dysplasia with PVP. The patient continued to have stable pain relief and neurologic improvement with independence of all daily living activities at 1-year follow-up.

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