

In a Seemingly Silent and Empty Desert, A Lion might be Hidden

Rahimizadeh Abolfazl, Habibi Gholam Reza, Rahimizadeh Ava

Pamim Research Center, Pars Hospital Neurosurgery Spine Tehran IR

ABSTRACT

Vertebral compression fractures are a leading cause of disability and morbidity in the patients suffering osteoporosis. Pain and kyphosis due to progressive vertebral collapse are the consequences of osteoporotic fractures. Furthermore, these fractures may affect quality of life, physical function, mental health and survival. Pain, which is the cardinal feature of such fracture, is incapacitating in the majority of the patients. In a number of the patients pain subsides within two to four weeks with proper immobilization. But, it may continue despite wearing orthosis.

Vertebroplasty and kyphoplasty have developed for substantial relief of pain and preserving function. However, kyphoplasty is superior to vertebroplasty because it has the benefits of restoring height and correcting the deformity.

Herein, we present a 39-year-old female who sustained osteoporotic compression fracture of the second lumbar vertebra following fall from the stairs. This patient had different story with time which has been not reported previously.

KEY WORDS: Kyphoplasty, lumbar spine, myxopapillary ependymoma, vertebroplasty

INTRODUCTION

The prevalence of osteoporosis increases with the increasing age of the population. However, in some countries, this type of fracture may occur at earlier age due to low calcium intake and vitamin D deficiency. Nonetheless, Osteoporotic vertebral compression fractures (OVCF) at any age is the major health problem worldwide. Percutaneous vertebroplasty (PVP) and percutaneous balloon kyphoplasty (PKP) can increase bone strength as well as alleviate the pain caused by vertebral compression fractures (VCFs), and both procedures rely on polymethyl methacrylate (PMMA) cement injected into the fractured vertebra for mechanical stabilization of the VCFs.

In the spinal cord, ependymomas represent approximately 63% of all parenchymal tumors, although figures have ranged from 30% to 88%. Myxopapillary ependymoma (MEPNs) commonly affects the filum terminale and represents 13% of all ependymomas. These tumors might be silent. Low back pain is the first complaint of the patient

and aggravates during sleeping hours. This is gradually associated with urinary disturbances and lower limb weakness. They are usually well-circumscribed lesions, making complete removal possible in most cases.

Herein we present a woman who was referred to our institute with the diagnosis of L2 osteoporotic fracture. Review of her radiographs revealed L2 compression fracture with affection of the superior end plate that was confirmed in reconstructed CT images. Despite an appropriate brace, analgesics and NSAIDs, the pain continued and some neural impairment was also gradually added. MRI was eventually performed and revealed a filum terminale tumor extending from L1 to L5. Complete removal of the tumor that was later diagnosed to be Myxopapillary ependymoma resulted in complete recovery of the patient.

CASE REPORT

A 39-year-old female was referred to our institute for kyphoplasty. She had history of falling from the

stairs, which subsequently resulted in back pain. She was transferred to a regional hospital where plain radiographs revealed L2 osteoporotic fracture (Figure 1A, B). Bone mineral densitometry showed osteoporosis, probably being attributed to her premature ovarian failure (POF). Serum D3 was 9.5 where Ca, P and PTH were normal. Substantial immobilization in an appropriate brace was recommended and was used correctly by this compliant patient. Calcium plus vitamin D, Strontium and Calcitonin were also prescribed.

Pain was initially decreased with the orthosis, but gradually became intolerable again. This was followed with mild paresthesia in the lower limbs. Therefore, reformatted CT scan was done in that hospital which confirmed osteoporotic compression fracture, but without canal compromise (Figure 2A, B). Therefore, kyphoplasty was recommended and she was transferred to our institute.

On admission to our hospital, her neurological examination was normal. With consideration of the normal neurology, we also agreed that she should undergo kyphoplasty. The night before the procedure, T1- and T2-weighted MRI as well as STIR were recommended and performed. Surprisingly, this revealed a cauda equina tumor extending from L1 to S1 (Figure 3A, B). GD MRI showed enhancement of the tumor (Figure 4).

With a clinical diagnosis of myxopapillary ependymoma, the patient underwent surgery the following day. Under general anesthesia and with the patient in the prone position, laminectomy from L1 to L5 was done. Subsequently and after dural opening, an encapsulated tumor extending from L1 to L5 was seen. With the possibility of myxopapillary ependymoma, en block total removal of the tumor was achieved (Figure 5A, B). However, despite the wide laminectomy, instrumentation was not done because of

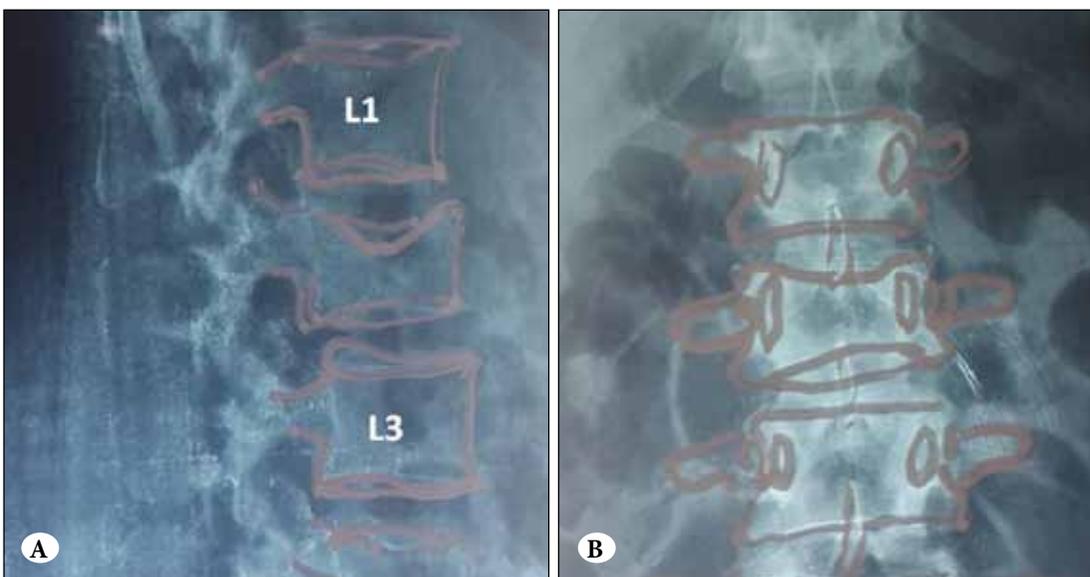


Figure 1: Lumbar plain radiographs: A, B) lateral and AP views showing compression fracture of second lumbar vertebra. The superior end plate is collapsed.



Figure 2: A,B) sagittal and coronal reformatted CT scan of the lumbar spine confirming L2 compression fracture.



Figure 3: MRI of the lumbar spine. **A)** T1-weighted image showing a cystic mass being started from T12 to L2, from L2 to L5 is isointense. **B)** T2-weighted MRI demonstrating L2 compression fracture and a cystic mass extending from T12 to L2 and from L2 to L5 is suspicious of a mass. **C)** STIR showing L2 fracture with edema at the superior part of the vertebra; the entire lumbar region is hyperintense.



Figure 4: GD MRI: shows marked enhancement of a mass from L2 to L5.

the refusal of her family. The postoperative course was uneventful and she was discharged a few days later, in an orthosis with her previous back pain dramatically decreased in severity. At the 6-month follow up, she was doing well and the lumbar spine standing radiograph did not show a deformity, although she was strongly advised to undergo periodic X-ray evaluations.

DISCUSSION

Osteoporotic vertebral fractures constitute a major health problem for the patients, resulting in worse quality of life compared to the normal population (4). Vertebroplasty (VP) and balloon kyphoplasty (KP) are less invasive procedures to treat such fractures (2,9). Both of these procedures can produce immediate pain relief compared to conservative treatments. Functional improvement is the superiority of these procedures. However, kyphoplasty has a significant advantage over vertebroplasty in terms of kyphosis correction, vertebral height restoration, and cement leakage prevention (2,4,9). The first line of treatment in osteoporotic compression fractures consists of sustained application of orthosis, partial bed rest, non-steroidal anti-inflammatory drugs (NSAIDs) and painkillers from paracetamol to narcotics. If a patient fails to improve in terms of pain, vertebroplasty or kyphoplasty are indicated (2,4,9).

On the other hand, myxopapillary ependymoma is defined as a slow-growing glioma and is considered a distinct variant of ependymomas classified as World Health Organization grade I.

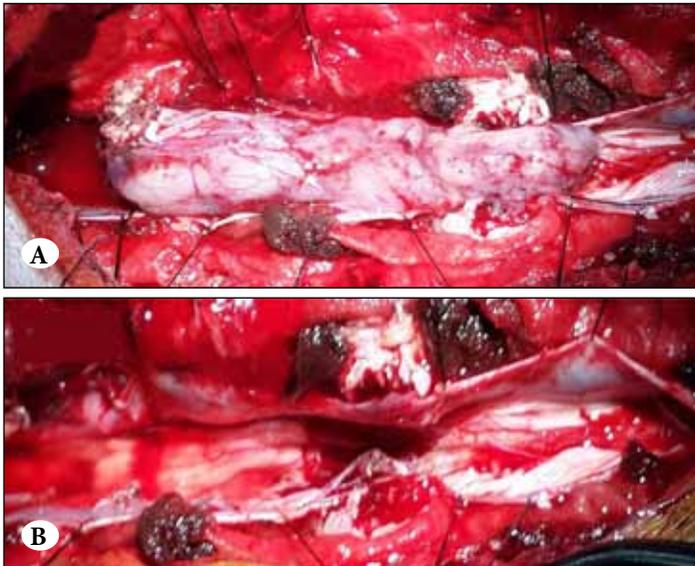


Figure 5: A) Intraoperative photograph: shows a huge mass. B) Note the integrity of the cauda equina after tumor removal.



Figure 6: The size of the tumor compared to a surgical knife.

Myxopapillary ependymoma is a distinctive subtype of glioma that occurs almost exclusively in the filum terminale and conus medullaris, accounting for nearly 90% of primary tumors in the cauda equina region. Its annual incidence is less than 0.01% per 100,000. Most arise from the filum terminale and are therefore mainly located at the cauda equina with occasional extension into the conus medullaris. At the filum terminale, these ependymomas are thought to arise from the ependymal rests ectopically deposited during development. Less frequently, myxopapillary might arise directly in the conus (10,11).

Myxopapillary ependymomas tend to occur in younger patients and are more prevalent in men. They typically present in the third to fifth decade of life. The average age at diagnosis is around 40 years (10,11).

Myxopapillary ependymomas tend to have an insidious presentation with gradual onset of the symptoms and prolonged history. They usually present with slowly progressive low back pain (5,10,11). Sensory deficits presenting as paresthesia of the lower limbs usually occur very late (5,6, 10,11). This might be rarely associated with subsequent involvement of the sphincter and sexual functions (10,11). The interval between the appearance of low back pain and the diagnosis varies from one month to 10 years with mean value of 18 months.

Gross total resection (GTR) is the gold standard in surgical treatment of myxopapillary ependymomas (5). Therefore, the surgeon should be cautious not to violate the capsule. If the capsule is violated, dissemination via cerebrospinal fluid will be possible (1). In terms of local recurrence, the younger the patient at the time of diagnosis (3,7-8,12). In terms of post surgery recurrence, shorter duration of symptoms before surgery and incomplete resection are closely related to the likelihood of recurrence (3,7,8,12).

Actually, these tumors tend to disseminate and recur despite their benign histologic character. Therefore, post-operative radiotherapy (RT) has been advocated as an additional control measure for patients with subtotal resection (3). Some authors have proposed whole brain, spinal cord radiotherapy in addition to local radiotherapy (1).

Compared with intracranial ependymomas, myxopapillary ependymomas have a better prognosis (3,7,8). Nonetheless, total removal of the tumor and the absence or the presence of tumor cell at the surgical margins of the initial surgery are crucial factors in determining the prognosis. Some long-term follow-up studies have suggested that recurrence rates may be higher than originally thought. However, 5-year survival rates are quoted as being up to 92% (12). Whole brain and spinal canal radiotherapy carries better overall prognosis in those with subtotal removal (1).

In conclusion, the current case is a good example of a medical curiosity that might be hidden in a seemingly simple condition. “In a seemingly silent and empty desert, a lion might be hidden”

REFERENCES

1. Fassett D, Pingree J, Kestle AW. The high incidence of tumor dissemination in myxopapillary ependymoma in pediatric patients. *J Neurosurg.* 102: 59–64,2005.
2. Lieberman IH, Dudeney S, Reinhardt MK, Bell G. Initial outcome and efficacy of “kyphoplasty” in the treatment of painful osteoporotic vertebral compression fractures. *Spine.* 26:1631-1638,2001.

3. Lin YH, Huang CI, Wong TT, Chen MH, Shiau CY, Wang LW, Ming-Tak Ho D, Yen SH. Treatment of spinal cord ependymomas by surgery with or without postoperative radiotherapy. *J Neurooncol.* 71:205–210,2005.
4. Liu JT, Liao WJ, Tan WC, Lee JK, Liu CH, Chen YH, TB. L. Balloon kyphoplasty versus vertebroplasty for treatment of osteoporotic vertebral compression fracture: A prospective, comparative, and randomized clinical study. *Osteoporos Int.* 21:359-364,2010.
5. McCormick PC. Microsurgical en-block resection of myxopapillary cauda equina ependymomas. *Neurosurg Focus.* 37: Suppl 2,2014.
6. Mercer C. A case study of spinal ependymoma presenting as non-resolving back and leg pain, Case report. *Manual Therapy.* 15: e603-606,2010.
7. Nakamura M, Ishii K, Watanabe K et al. Long-term surgical outcomes for myxopapillary ependymomas of the cauda equina. *Spine.* 34: E756–760, 2009.
8. Pica A, Miller R, Villa S, et al. The results of surgery, with or without radiotherapy, for primary spinal myxopapillary ependymoma: A retrospective study from the rare cancer network. *Int J Radiat Oncol Biol Phys.* 74:1114-1120, 2009.
9. Phillips FM. Minimally invasive treatments of osteoporotic vertebral compression fractures. *Spine.* 28:S45-S53,2003.
10. Schweitzer JS, Batzdorf U. Ependymoma of the cauda equina region: Diagnosis, treatment, and outcome in 15 patients. *Neurosurgery.* 1992;30(2):202e7.
11. Sonneland PR, Scheithauer BW, Onofrio BM. Myxopapillary ependymoma. *Cancer.* 56:883–93,1985.
12. Zaucha R, Sosinska-Mielcarek K, Zander I, Szewczyk P, Jassem J. Spinal cord ependymoma: Retrospective analysis of treatment outcomes of 14 patients. *J Radiotherapy in Practice.* 4:155:e60-66,2004.

Manuscript submitted July 23, 2016.

Accepted February 09, 2017.

Address correspondence to: Abolfazl Rahimizadeh, Pars Spine Institute and PAMIM Research Center, Pars Hospital, Iran University of Medical Sciences, Tehran, Iran, 67 Keshwarz Blvd,1415944911Tehran, Islamic republic of Iran

Phone: +98 218 866 12 44

email: a_rahimizadeh@hotmail.com