Case Report | Journal of Orthopaedic Case Reports 2022 October:12(10):Page 34-38

Strategies in the Management of Osteoporotic Kummell's Disease

Naveenkumar Kuppan[1], Sathish Muthu[2,3], Sathyanarayanan Parthasarathy[1], Pragash Mohanen[1]

Learning Point of the Article:

Osteoporotic Kummell's disease needs careful planning of management strategies since they are more prone to pseudoarthrosis due to poor vascular and mechanical support.

Abstract

Introduction: Kummell disease is a condition characterized by severe pain, progressing kyphosis with or without neurological deficit following a trivial trauma in the old age osteoporotic population. It is an osteoporotic vertebral fracture due to avascular necrosis of the vertebra, having an asymptomatic period initially followed by progressive pain, kyphosis, and neurologic deficit. Although various management options are available for Kummell's disease, a dilemma occurs in selecting an optimal modality in each case.

Case Report: A 65-year-old female presented with complaints of low back pain for 4 weeks. She developed progressive weakness and bowel bladder disturbance. Radiographs showed a D12 vertebral compression fracture with an intravertebral vacuum cleft sign. Magnetic resonance imaging showed intravertebral fluid and significant compression of the cord. We performed posterior decompression, stabilization, and transpedicular bone grafting at the D12 level. Histopathology confirmed Kummell's disease. The patient recovered with restored power and bladder control and resumed independent ambulation.

Conclusion: Osteoporotic compression fractures are more prone to pseudoarthrosis due to poor vascular and mechanical support, they need adequate immobilization and bracing. Transpedicular bone grafting for kummels disease seems to be a good surgical option due to its short operating time, less bleeding, less invasive approach, and early recovery. However, a treatment-oriented classification is needed to treat this clinical entity on a case-by-case basis.

Keywords: Kummell's disease, osteoporosis, kyphoplasty, transpedicular bone grafting, spine.

Introduction

Kummell's disease is an osteoporotic vertebral fracture due to avascular necrosis of the vertebra, having an asymptomatic period initially followed by progressive pain, kyphosis, and neurologic deficit [1]. It is a condition characterized by severe pain and progressive kyphosis with or without neurological deficit following a trivial trauma in the old age osteoporotic population [2]. It is also known as delayed post-traumatic osteonecrosis and is rarely reported in the literature. First described by Dr. Hermann Kummell in 1891, it poses a diagnostic challenge and when missed, causes life-threatening complications including acute spinal cord compression [3, 4]. Kummel's disease is a diagnosis of exclusion based on clinical history and radiological and histopathological findings [5]. Various management options are available for Kummell's disease ranging from conventional decompression and fusion to minimally invasive strategies such as kyphoplasty [6, 7]. A dilemma occurs in selecting an optimal modality in each case.

We report a case of Kummell's disease in an old age women, with its diagnostic challenges and surgical management. We also present the

Access this article online

Website: www.jocr.co.in

DOI: 10.13107/jocr.2022.v12.i10.3356

Author's Photo Gallery


Dr. Naveenkumar Kuppan


Dr. Sathish Muthu


Dr. Sathyanarayanan Parthasarathy


Dr. Pragash Mohanen

[1]Department of Orthopaedics and Spine surgery, Sree Manakula Vinayagar Medical College and Hospitals, Puducherry, India.
[2]Department of Orthopaedics, Government Medical College, Dindigul, Tamil Nadu, India,
[3]Orthopaedic Research Group, Coimbatore, Tamil Nadu, India.

Address of Correspondence:
Dr. Sathish Muthu,
Department of Orthopaedics, Government Medical College, Dindigul, Tamil Nadu, India.
E-mail: drsathishmuthu@gmail.com

Submitted: 19/06/2022; Review: 01/08/2022; Accepted: September 2022; Published: October 2022

DOI:10.13107/jocr.2022.v12.i10.3356

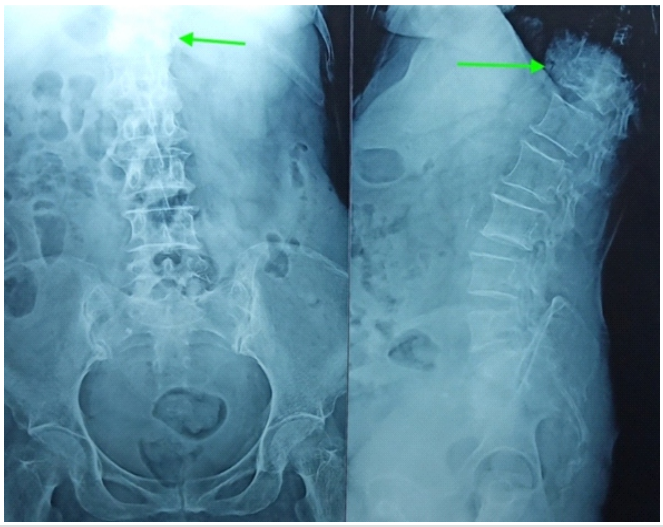

Figure 1: Pre-operative radiological presentation of the patient with green arrows showing the level of compression fracture at D12.

strategies to be employed in the management of such disease when compounded in an osteoporotic scenario which is more prone to pseudoarthrosis.

Case Presentation

A woman in her seventh decade presented with complaints of low back pain for 4 weeks. She had difficulty walking and standing due to progressive weakness and bowel bladder disturbance for 4 weeks. She complained of loss of sensation in both lower limbs for the past 4 weeks. She alleged to have fallen at her home 2 months ago. She was treated conservatively with a brace and medications. She developed progressive weakness after 4 weeks following the injury. There was no history of fever, evening rise of temperature, sudden weight loss, or loss of appetite. On examination, there was diffuse spine tenderness noted over the dorsolumbar region, localized kyphosis with paraspinal muscle stiffness. The neurological examination of the patient is mentioned in (Table 1).

Radiographs revealed a D12 vertebral compression fracture with an intravertebral vacuum cleft sign (Fig. 1). Magnetic resonance imaging (MRI) showed increased signal intensity within the D12 vertebra showing intravertebral fluid, a linear area of decreased intensity on either side representing intravertebral vacuum cleft (double line sign), and significant compression of the cord at the level (Fig. 2). There was no paravertebral fluid or mass noted. The patient was diagnosed to have a D12 pathological anterior wedge compression fracture with paraparesis.

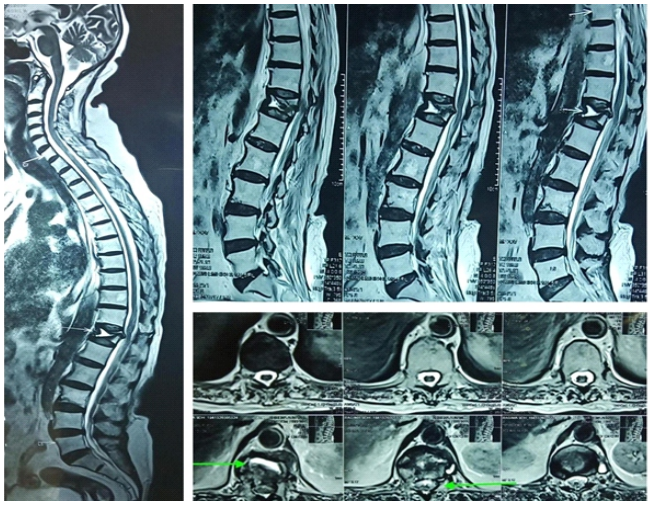

Figure 2: Magnetic resonance imaging showing increased signal intensity within D12 vertebra showing intravertebral fluid (white arrow), a linear area of decreased intensity on either side representing intravertebral vacuum cleft (double line sign), and significant compression of the cord at the level (green arrow).

We considered differential diagnoses such as osteoporotic wedge compression fracture, post-traumatic kyphosis, Potts's spine, metastasis, hemangioma, and osteonecrosis of the spine. Pre-operative investigations including total count, leukocyte count, erythrocyte sedimentation rate, C-reactive protein, serum Vitamin D, serum electrophoresis, and serum alkaline phosphatase were all normal. DEXA scan diagnosed the patient to have osteoporosis with T score of −2.7 and Z score of −2.2. Computed tomography (CT) chest and abdomen showed no mass or abnormality.

A provisional diagnosis of pseudoarthrosis of the spine (Kummell's disease) was arrived upon excluding all other differential diagnoses. We proceeded with posterior decompression, stabilization, and transpedicular bone grafting at the D12 level. The manual reduction was obtained by prone positioning the patient. Intraoperatively, decompression of the cord was done at the D12 level. Transpedicular biopsy was done through the left D12 pedicle which showed bony fragments, no pus or abnormal tissues were

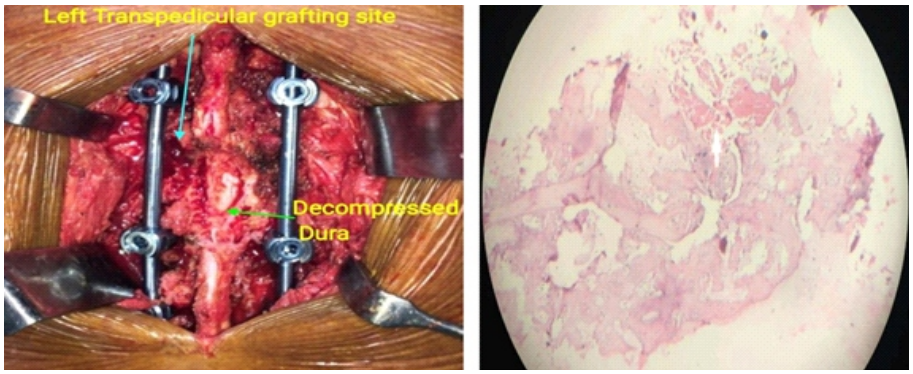

Figure 3: Intraoperative image showing the decompressed dura and the site of transpedicular biopsy without any pus or abnormal tissues (a). A histological image of the biopsied specimen shows necrotic bony fragments with occasional hematopoietic elements present suggestive of sequestrum noted in Kummell's disease (b).

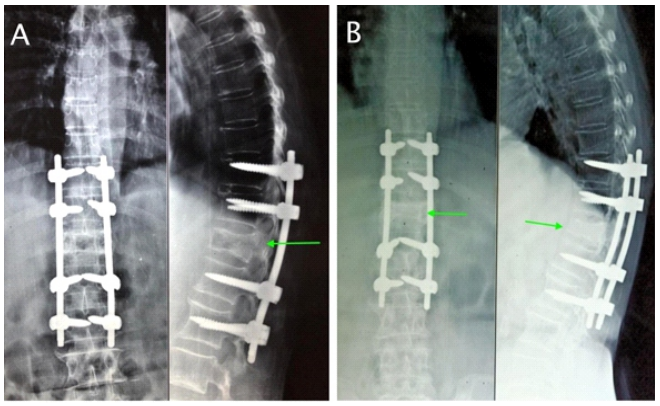

Figure 4: (a) The immediate decompression and stabilization of the compression fracture with reconstitution of the vertebral height with bone grafting (green arrow) and (b) the consolidation of the graft with the vertebral body (green arrow) at 1-year follow-up.

present (Fig. 3). Hence, we proceeded with transpedicular vertebral bone grafting of the D12 vertebra through a left-sided pedicle using a local bone graft. Posterior long segment stabilization was done from D10 to L2 levels (Fig. 4a). Biopsy specimens were sent for microbiological and histopathological studies. Gram staining and acid-fast bacilli staining were negative. The culture showed no growth of aerobes or anaerobes. The nucleic acid amplification test for Mycobacterium tuberculosis was also negative. Histopathology report showed necrotic bony fragments with occasional hematopoietic elements present (Fig. 3), features suggestive of sequestrum, confirming the diagnosis of Kummell's disease.

Postoperatively, the patient was mobilized from day 2. Osteoporosis was managed with daily subcutaneous injection of 20 μg of recombinant human parathyroid hormone analog along with supplemental calcium and Vitamin D3. Significant improvement in motor power was noted in bilateral lower limbs from day 3. Neurological status at 3- and 12-month follow-up showed significant recovery as in Table 2. At 1-year follow-up, the patient was ambulant independently and radiographs show good graft uptake (Fig. 4b).

Table 1: Pre-operative neurological assessment of the patient

Side	Right	Left
Tone	Decreased	Decreased
Bulk	Wasted	Wasted
L2	3/5	0/5
L3	2/5	0/5
L4	1/5	0/5
L5	1/5	0/5
S1	1/5	0/5
Sensory	Decreased below L1	Decreased below L1

Discussion

Kummell's disease is avascular necrosis of the vertebral body causing delayed vertebral collapse following trivial trauma [1]. The usual presentation is backache followed by progressive kyphosis. The prevalence in osteoporotic compression fractures ranges from 7% to 37% [2], with a higher incidence in men. Radiographic presentations are vertebral compression fracture non-union, vacuum defects in the intervertebral region, delayed collapse of vertebral body, osteonecrosis of vertebral body post-trauma, or pseudo arthrosis of vertebral body. The proposed theories of pathology include mechanical theory due to loss of nutritional contribution resulting in fragility and bone collapse and vascular theory due to watershed zone of the anterior third of vertebral body resulting in bone collapse [3]. Predisposing conditions include atherosclerosis, hemoglobinopathies, vasculitis, cirrhosis, Gaucher's disease, and sarcoidosis. It commonly involves the thoracolumbar region from T8 to L4 in 60 % of cases.

In most cases, initial trauma shows no significant radiological finding with an asymptomatic period ranging from months to years, followed by severe pain without any trauma, progressive kyphosis, and new onset neurological deficit. Subsequent radiological examination shows vertebral body collapse, vacuum cleft, vertebral osteonecrosis, and transverse or linear radiolucent shadows in the vertebral body endplates. Dynamic radiographs show a change in vacuum cleft size. CT is the earliest modality to find micro fractures and intravertebral vacuum cleft signs. The vacuum cleft phenomenon (Kummell's sign) is also seen in other conditions such as malignancy, but a linear-shaped vacuum cleft is definitive of benign lesions such as osteonecrosis [4]. The features to distinguish between a benign osteoporotic vertebral body collapse and malignant vertebral body collapse are listed in Table 3 [5].

MRI shows a necrotic region with increased signal intensity on T1 and decreased signal intensity on T2. The intravertebral fluid shows hyperintensity in the T2 image surrounded by a region of decreased signal intensity on either side showing a "Double line sign" [6]. Kummel's disease is a diagnosis of exclusion based on the

Table 2: Post-operative neurological improvement of the patient at 3- and 12-month follow-up period

Follow-up	3 months		1 year	
Side	Right	Left	Right	Left
Tone	Normal	Normal	Normal	Normal
L2	4/5	4/5	5/5	5/5
L3	4/5	4/5	5/5	5/5
L4	4/5	3/5	5/5	5/5
L5	4/5	3/5	5/5	4/5
S1	4/5	3/5	5/5	4/5
Sensory	Recovered	Recovered	Intact	Intact

Table 3: Comparative features to distinguish between a benign osteoporotic vertebral body collapse and malignant vertebral body collapse

Modality	Benign collapse	Malignant collapse
MRI: morphology	Normal signals in posterior elements with retropulsion of bone fragments into the spinal canal and additional benign vertebral collapse.	Abnormal signals in posterior elements with epidural and paravertebral soft-tissue mass, expanded posterior vertebral contour, and metastasis in additional vertebrae.
MRI: signal intensity and contrast enhancement patterns	Normal bone marrow signal preserved, margins regular, linear horizontal hypointense T1/T2 band, intravertebral cleft with fluid sign, double-line sign, and normal enhancement relative to adjacent vertebrae.	Bone marrow signal geographically replaced, margins irregular, increased enhancement relative to adjacent vertebrae.
MRI: diffusion pattern	Diffusion not restricted.	Restricted diffusion pattern.
CT	Retropulsion of bone fragments, puzzle sign, sharp vertebral fracture lines, and intravertebral vacuum phenomenon.	Destruction of bone with epidural or focal paravertebral soft-tissue mass

clinical history and radiological and histopathological findings. Omidi-Kashani et al. [7] reported the diagnostic criteria for Kummell's disease based on the clinical course of trivial trauma with an asymptomatic period followed by progressive symptoms refractory to conservative measures, progressive vertebral body collapse, intravertebral vacuum cleft sign, double line sign, and necrotic bone fragments in histopathology.

Steel et al. [8], classified Kummell's disease into five stages based on clinical findings: Stage 1, traumatic stage without obvious abnormalities, Stage 2, asymptomatic stage, Stage 3, subclinical period with minor symptoms, Stage 4, gradually worsening back pain, and Stage 5, with kyphotic deformity and cord compression. Li et al. [9], did an MRI-based staging of Kummell's disease into three stages: Stage 1 Height loss of vertebral body <20%, Stage 2 Height loss of vertebral body >20% with adjacent disc degeneration, and Stage 3 with posterior body involvement and cord compression.

Management involves absolute bed rest, analgesics, bracing, lumbar traction, and immobilization. Surgery is indicated in cases with severe pain, progressive worsening of kyphosis, and worsening neurology [10]. Surgical options involve anterior/posterior decompression and fusion and minimally invasive procedures such as vertebroplasty or kyphoplasty [11]. In patients without neurological symptoms, procedures such as percutaneous vertebroplasty (PVP) and percutaneous kyphoplasty (PKP) are found to be effective in relieving the pain and restoration of the vertebral body height [12]. In patients presenting late especially in Stage 3, PVP or PKP are less safe and less effective, as there is a break in the posterior vertebral cortex with a risk of cement leakage into the spinal canal which also worsens the intravertebral instability as noted in our case. Therefore, other options such as stabilization and fusion, transpedicular bone grafting [13] or augments (titanium mesh bone grafting) [14, 15], or osteotomy are the alternative measures.

Li et al. [9], based on his MRI-based staging system advocated augmentation procedures such as vertebroplasty or kyphoplasty in Stage 1 and 2 diseases, and decompression with fusion for Stage 3 disease. Kim et al. [16], stated that in cases with canal compromise and neurologic deficit, decompression with stabilization and fusion is indicated. Li et al. [17], stated transpedicular insertion of vertebral augments combined with short segment fixation yielded good results in cases of canal compromise. Chen et al. [10], in their study on 54 patients found that there is no difference based on clinical outcome for cases undergone kyphoplasty alone versus short segment fixation with vertebroplasty. Yu et al. [18], reported no use of a balloon for further expanding fractured vertebra, while postural reduction can effectively correct kyphosis and cord compression. Li et al. [17], in their study on 21 patients with Stage 3 Kummell's disease managed using short segment fixation and transpedicular body augmenter, found excellent results and less complication rate. Wang et al. [19] in his study on the treatment of 30 patients with Kummell's disease, compared between anterior approach and transpedicular intracorporeal grafting with posterior stabilization and found no statistical difference in terms of post-operative VAS grade, union, operative time, and kyphosis correction angle. They reported that posterior operation can effectively restore anterior vertebral height and correct kyphosis similar to the anterior approach, with reduced surgical morbidity compared the anterior approach. In a minimally invasive perspective, attempts were made to reconstruct the anterior column by transpedicular intracorporeal cage grafting with short-segment fixation thereby preventing violation of the paravertebral musculature to the compromised spine segment posteriorly [20,21].

A novel vertebral body osteonecrosis classification [22] was proposed to grade the disease based on the relevant imaging findings and sagittal biomechanical parameters. However, the classification dose not guide the selection of the appropriate treatment option from the list of treatment methods available for managing this disease. Hence, there is a need for a treatment-oriented classification to better understand and treat this clinical entity.

Conclusion

Osteoporotic compression fractures are more prone to pseudoarthrosis due to poor vascular and mechanical support and

they need adequate immobilization and bracing. Transpedicular bone grafting for kummels disease seems to be a good surgical option due to its short operating time, less bleeding, less invasive approach, and early recovery. However, a treatment-oriented classification is needed to treat this clinical entity on a case-by-case basis.

Clinical Message

Osteoporotic compression fractures are more prone to pseudoarthrosis due to poor vascular and mechanical support, and they need adequate immobilization and bracing. Kummell's disease should be considered as a differential for back pain in the elderly, as early diagnosis helps to prevent complications. Transpedicular bone grafting for Kummell's disease seems to be a good surgical option due to the short operating time, less bleeding, less invasive approach, and early recovery. A thorough knowledge of the disease pathology and a high level of suspicion is needed for early diagnosis. There is a need for treatment-oriented classification to better understand and treat this clinical entity.

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

Conflict of interest: Nil **Source of support:** None

References

1. D'Oria S, Delvecchio C, Dibenedetto M, Zizza F, Somma C. Case report of Kummell's disease with delayed onset myelopathy and the literature review. Eur J Orthop Surg Traumatol 2018;28:309-16.
2. Freedman BA, Heller JG. Kummel disease: A not-so-rare complication of osteoporotic vertebral compression fractures. J Am Board Fam Med 2009;22:75-8.
3. Matzaroglou C, Georgiou CS, Panagopoulos A, Assimakopoulos K, Wilke HJ, Habermann B, et al. Kummell's disease: Clarifying the mechanisms and patients' inclusion criteria. Open Orthop J 2014;8:288-97.
4. Bhalla S, Reinus WR. The linear intravertebral vacuum: A sign of benign vertebral collapse. AJR Am J Roentgenol 1998;170:1563-9.
5. Mauch JT, Carr CM, Cloft H, Diehn FE. Review of the imaging features of benign osteoporotic and malignant vertebral compression fractures. AJNR Am J Neuroradiol 2018;39:1584-92.
6. Huang YS, Hao YS, Wang XD, Sun HH, Du JP, Yang JS. Long-segment or bone cement-augmented short-segment fixation for Kummell disease with neurologic deficit? A comparative cohort study. World Neurosurg 2018;116:e1079-86.
7. Omidi-Kashani F, Parsa A, Madarshahian D. Impending cauda equina syndrome due to Kummell disease; A case report and literature review. Int J Surg Case Rep 2021;83:106041.
8. Steel HH. Kummell's disease. Am J Surg 1951;81:161-7.
9. Li KC, Wong TU, Kung FC, Li A, Hsieh CH. Staging of Kummell's disease. J Musculoskelet Res 2004;8:43-55.
10. Chen L, Dong R, Gu Y, FengY. Comparison between balloon kyphoplasty and short segmental fixation combined with vertebroplasty in the treatment of Kümmell's disease. Pain Physician 2015;18:373-81.
11. Jiang J, Gu FL, Li ZW, Zhou Y. The clinical efficacy and experience of bipedicular percutaneous vertebroplasty combined with postural reduction in the treatment of Kümmell's disease. BMC Musculoskelet Disord 2020;21:82.
12. Zhang J, Fan Y, He X, Meng Y, Huang Y, Jia S, et al. Is percutaneous kyphoplasty the better choice for minimally invasive treatment of neurologically intact osteoporotic Kummell's disease? A comparison of two minimally invasive procedures. Int Orthop 2018;42:1321-6.
13. Zhu Y, Zhang Z, Jiang W, Su K, Wang Z, Wang C, et al. Therapeutic efficacy of transpedicular impaction bone grafting with long segmental posterior instrumentation in stage III Kümmell disease. Spine (Phila Pa 1976) 2021;46:907-14.
14. Mei L, Sang W, Chen Z, Lou C, Zheng L, Jin K, et al. Titanium mesh bone grafting combined with pedicle screw internal fixation for treatment of Ku [Combining Diaeresis] mmell disease with cord compression: A case report and literature review. Medicine (Baltimore) 2018;97:e12183.
15. Lee SH, Jain V, Lee SH, Baek OK, Moon KH. Transpedicular intrabody cage insertion with posterior spine stabilization in Kümmell disease: Report of 2 cases. World Neurosurg 2018;116:236-40.
16. Kim KT, Suk KS, Kim JM, Lee SH. Delayed vertebral collapse with neurological deficits secondary to osteoporosis. Int Orthop 2003;27:65-9.
17. Li KC, Li AF, Hsieh CH, Liao TH, Chen CH. Another option to treat Kummell's disease with cord compression. Eur Spine J 2007;16:1479-87.
18. Yu HM, Li YZ, Yao XD, Lin JK, Pan YC, Zhuang HF, et al. Surgical options for delayed osteoporotic vertebral collapse. Zhongguo Gu Shang 2016;29:606-13.
19. Wang F, Wang D, Tan B, Dong J, Feng R, Yuan Z, et al. Comparative study of modified posterior operation to treat Kummell's disease. Medicine (Baltimore) 2015;94:e1595.
20. Park HJ, Kim HB, You KH, Kang MS. Percutaneous transpedicular intracorporeal cage grafting for Kümmell disease. Acta Neurochir (Wien) 2022;164:1891-94.
21. Chen C, Gao X, Li H, Pan X, Wang S. Intravertebral insertion of interbody fusion cage via transpedicular approach for the treatment of stage III Kümmell disease: A technical note and case presentation. Br J Neurosurg 2021;1-6.
22. Formica M, Zanirato A, Cavagnaro L, Basso M, Divano S, Formica C, et al. What is the current evidence on vertebral body osteonecrosis? A systematic review of the literature. Asian Spine J 2018;12:586-99.

Conflict of Interest: Nil
Source of Support: Nil

Consent: The authors confirm that informed consent was obtained from the patient for publication of this case report

How to Cite this Article

Kuppan N, Muthu S, Parthasarathy S, Mohanen P. Strategies in the Management of Osteoporotic Kummell's Disease. Journal of Orthopaedic Case Reports 2022 October;12(10): 34-38.