Kummell's Disease: A Case Report and Review of Literature

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The incidence of osteoporosis and osteoporotic vertebral compression fractures are increasing with increase in life expectancy. Most of the vertebral compression fractures heal without any complications but few may develop kyphotic deformity, persistent pain and even osteonecrosis or non-union of the fracture. The non-union or pseudoarthrosis of the osteoporotic vertebral compression fracture is called Kummell's disease which has its own characteristic clinical stages and radiological features.

We are reporting a case report of 82 years old lady that demonstrates typical clinical and radiological features of Kummell's disease. All the clinicians should be aware of this condition because not only orthopaedic or spine surgeons but other specialities also manage osteoporosis and osteoporotic vertebral compression fractures in their day to day practice.

Keywords: Kummel's Disease, osteoporosis, vertebral fracture.

The incidence of osteoporosis and osteoporotic vertebral compression fractures are increasing with increase in life expectancy. Most of the vertebral compression fractures are stable and heal without complications, very few progress to develop osteonecrosis which is also known as Kummell's disease. The incidence of Kummell's disease is difficult to accurately report because multiple synonymous terms have been used to describe this pathology: posttraumatic vertebral osteonecrosis, vertebral pseudoarthrosis, intravertebral vacuum cleft or gas, delayed vertebral collapse and vertebral compression fracture non-union.[1,2] Regardless of terminology, the true incidence is actually quite high (7% to

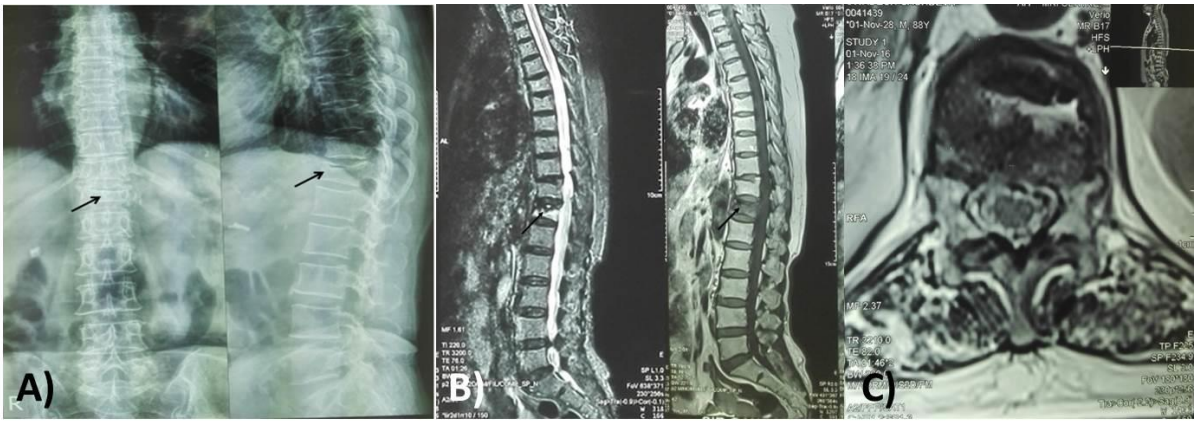

Figure 1: A) pre-operative X-ray showing intravertebral vacuum cleft in D12 vertebra, B) MRI T1 weightted and T2 weighted image showing "Fluid Sign", C) MRI Axial section showing involvement of anterior one third of the vertebral body

37%), especially in elderly population.

Kummell's disease usually occurs in elderly and middle aged population. Patient develops mild back pain after trivial trauma. There is usually asymptomatic period of weeks to months after which pain reappears and is severe. Sometimes patient may present with significant kyphotic deformity and neurological involvement. Radiography shows linear radiolucent shadow in the vertebral body or just underneath either end plates suggesting intraosseous vacuum cleft.[7] Magnetic Resonance Imaging (MRI) is an important imaging to rule out other pathologies and to see for a classic sign of osteonecrosis called "Fluid Sign".[3] The treatment options are conservative and surgical management with decompression and stabilization or vertebral augmentation procedures.

Although it was first described more than 100 years ago, difficulties with diagnosis and management of Kummell's disease still remain.[3,4]

We are reporting a case of 82 year old lady who sustained a trivial trauma 3 months back. She was asymptomatic after trauma but later developed severe mid back pain. Imaging demonstrated classic sign of Kummell's disease. This is a case report with review of literature which discusses historical background, aetiopathogenesis, clinical presentation, imaging and treatment options of Kummell's disease.

Case Report

Eighty two years old lady sustained a trivial trauma 3 months before reporting to us, while walking and sustained injury to her back. She was able to perform her daily activities with minimal pain for one and half months then developed severe pain in the same region. The pain was gradually progressive, localized, not associated with paresthesia or numbness. She visited local centre where she was advised conservative management with Taylor's Brace, analgesics and supplemental therapy. The pain worsened and she developed difficulty in walking. She came to our centre 3 months after the injury with severe pain and difficulty in walking. There is no of history of any long term steroid intake. On examination, there was significant

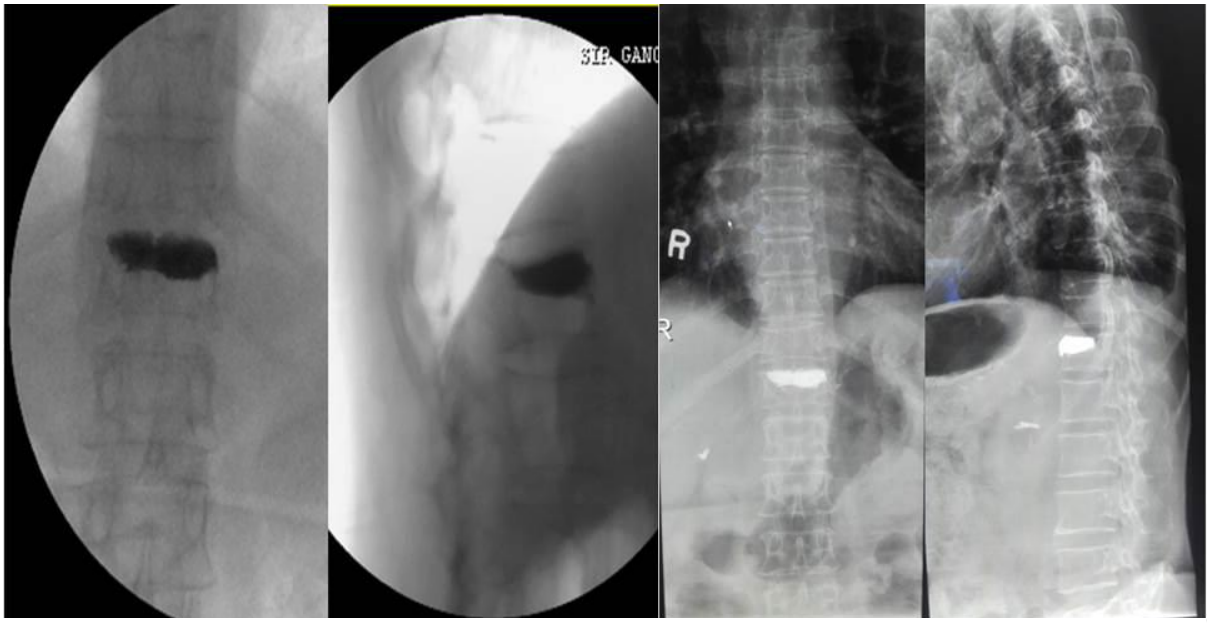

Figure 2: Post-operative X-ray after Percutaneous Balloon Kyphoplasty showing maintenance of vertebral body height

tenderness in dorsolumbar junction with kyphotic deformity. The neurological examination revealed normal findings.

Radiological evaluation of dorsolumbar spine with anteroposterior and lateral views revealed anterior wedging of D12 vertebra with linear radiolucency just underneath the superior endplate suggestive of intraosseous air. It also showed the healed fracture of D11 vertebra with anterior wedging **(Figure 1)**. The local kyphotic deformity was 17 degrees. MRI evaluation showed hypointensity in T1 weighted image and hyperintensity in T2 weighted image surrounded by hypointense ring shadow just underneath the superior endplate. The finding was consistent with fluid collection in a cavity which is also known as "Fluid Sign". There were no features suggestive of perivertebral inflammation.

The bone densitometry of the lumbar spine and hip showed T-score of -2.8 and -2.2 respectively. Haematological examination, biochemical evaluation and USG abdomen and pelvis did not reveal any significant findings suggestive of infection or malignancy.

As the patient did not respond to conservative management and the pain was so severe that it was affecting her daily activities; we performed percutaneous bipedicular balloon kyphoplasty. The preoperative anterior and middle height of the fractured vertebra improved from 64.7% and 55.55% of adjacent normal vertebral height to 84.61% and 83.33% respectively **(Figure 2)**. The kyphotic deformity improved significantly. She had significant pain relief postoperatively and was mobilized on the same evening with Taylor's brace and was discharged next day with anti-osteoporotic treatment. At 3 months follow up, the patient remained asymptomatic and radiographs revealed maintained vertebral height.

Discussion

Kummell's Disease was first described by German Surgeon Dr. Hermann Kummell in 1981.[5] He gave the description of this condition in six patients. According to him, this condition occurs in 3 stages; first stage is a stage of acute trauma followed by an asymptomatic period; second stage is a stage of recurrence of pain which is more severe than previous episode and third stage is appearance of kyphotic deformity and neurological deficit. This description was given prior to advent of radiography. Later in 1951 Steel elaborated this condition in 5 stages. First stage is an initial injury which varies in severity and mechanism but radiograph is necessarily negative. Second stage is post-traumatic period in which the patient may present with minor back pain without functional impairment. Third stage is during which patient is not incapacitated, may lasts for weeks to months. Fourth stage is recrudescent stage, the patient complains of persistent progressive back pain localized to the area of compressed fracture. Fifth stage when kyphotic deformity gradually occurs, with or without the development of spinal cord compression.

There is no consensus view regarding the aetiopathogenesis of Kummell's disease. A variety of hypothesis exists, including avascular osteonecrosis,[1,6] atrophic non-union,[7] microfracture, nutritional injury, pseudoarthrosis,[1] and fatigue fracture. The hypothesis developed by Kummell states that initial minor trauma to the vertebral body is not strong enough to cause fracture, but nutrition to vertebral body is damaged. The anterior one third of the vertebral body may represent a watershed zone due to the characteristics of blood supply in the region. Disruption of the watershed zone arterial supply leads to avascular necrosis mechanism for this entity. Osteonecrosis of the vertebral body has been associated with chronic steroid use, pancreatitis, sickle trait and Gaucher's disease. Chronic steroid use may lead to fatty infiltration of the vertebral body leading to compression of the intramedullary arteries compromising the vascular supply. Osteoporotic vertebral bodies are more prone to develop Kummell's disease due to distortion of trabecular microarchitecture and the consequent fragile nature of the bone. Low bone mass was found as a predisposing factor in about 55% of the reported cases.

The most common age group affected is elderly population, with a slight male predominance.[1] It usually affects the thoracolumbar junction, as it is a transition zone between rigid thoracic and mobile lumbar spine. There is usually a history of trivial trauma which leads to minimal pain which barely affects the daily activity of the patient. The patient is asymptomatic for few weeks to months; later develops pain which is more severe than the initial episode. The patient gradually develops kyphotic deformity and neurological symptoms, rarely involving bowel and bladder functions.

Ideally, Kummell's disease is diagnosed by serial plain X rays, in which initial films do not demonstrate evidence of a fracture. Maldague et al first described the intravertebral vacuum phenomenon. The intravertebral vacuum cleft is a linear,

transverse radiolucency due to accumulation of gas in the centre of the vertebral body or adjacent to one of the endplates.[7] It was thought that the intravertebral cleft is thought to be pathognomonic of Kummells disease but different authors have demonstrated the presence of intravertebral air in cases of malignancy, infection, osteoporosis, intraosseous disc prolapse.[8] Flexion and extension radiographs may be needed to detect pseudoarthrosis, which along with an intravertebral air filled cleft, are considered classic for Kummell's disease. CT scan better demonstrates the intravertebral vacuum cleft and has a heterogenous distribution and irregular shape when compared with X rays.

The appearance of the intravertebral vacuum cleft on MRI varies depending on whether it is filled with gas or fluid.[1,9] When filled with fluid, the cleft has a decreased signal on T1 weighted images and an increased signal on T2 weighted images.[10] This is called a "Fluid Sign" and is highly suggestive of avascular necrosis. A peripheral zone of hyperintensity surrounding hypointensity on T2 weighted image is called "Double Line Sign" and this corresponds to the intravertebral vacuum cleft. MRI scan should be obtained in all cases to rule out perivertebral inflammatory changes. In a study by Yu CW et al, vertebral collapse was significantly more severe in those having only intravertebral air than in those having intravertebral fluid with or without air.

There is no standard treatment and no single effective treatment for Kummel's disease.[11] Several factors must be taken into account when determining treatment modality, including the severity of pain, presence of neurological deficit, comorbidities, magnitude of the kyphotic deformity and bone quality.[1,9,12]

Non-surgical treatment options for Kummell's disease include bed rest, lumbar traction, wearing a brace, analgesics and anti-osteoporotic drugs. However the efficacy of treatment with brace and analgesics only is debatable.

In general, surgery is recommended for patients with serious pain refractory to conservative treatment, radiculopathy and/or increasing neurological deficit, or significant deformity. The aim of surgery is neural decompression and restoration of the normal sagittal balance of the spinal column.[11] Surgical interventions for Kummell's disease can be decided on the basis of Li's staging of the disease. According to Li et al Stage I is vertebral body compression <20%, Stage II is vertebral body compression >20% with adjacent disc involvement and stage III is vertebral body compression with posterior cortex involvement with spinal cord compression. They advocate augmentation alone in stage I and II and decompression with surgical stabilization in stage III. The vertebral augmentation can be done by percutaneous vertebroplasty or kyphoplasty which provides significant pain relief, improves function and corrects deformity.[1] Decompression with stabilization should be performed if there is any neurological deficit. The choice between an anterior and posterior procedure is based on the

surgeon's discretion; but in most cases, a both anterior and posterior fusion is required due to the presence of concomitant osteoporosis.

Conclusion

Kummell's Disease occurs in elderly and middle age patients with osteoporosis. The patients are usually asymptomatic after trivial trauma for weeks to months then gradually develop pain which is more severe and is associated with progressive kyphotic deformity and neurological symptoms. Transverse linear radiolucent sign suggestive of intraosseous air is a classic radiological sign. The treatment options are non-surgical, surgical decompression and stabilization and vertebral augmentation. Since most of these patients present to general physicians and orthopaedic surgeons in the initial stage, early identification and treatment can prevent the progression of the deformity and development of neurological deficit.

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