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Current Research Status and Progress in Kümmell's Disease

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Abstract: With the arrival of an aging society, the incidence of Kümmell's disease (KD), one of the complications of osteoporotic vertebral compression fracture (OVCF), has gradually increased, because it easily leads to vertebral instability and causes intractable pain or neurological dysfunction, and the disability rate of the disease is high, placing a serious burden on society and families. Since Dr. Kümmell Hermann first reported Kümmell's disease in 1895, more than 100 years later, the pathogenesis of this disease has not been elucidated, but many scholars generally believe that the presence of intravertebral vacuum cleft (IVC) sign on imaging is a specific sign of vertebral ischemic osteonecrosis and Kümmell's disease should be highly suspected. About two-thirds of patients with Kümmell's disease have no significant back pain symptoms in the early stages, resulting in patients rarely going to the hospital for relevant examinations. At the same time, trivial injuries are easily overlooked by patients, so it is difficult to make an early diagnosis of this disease. Progressive kyphosis due to vertebral collapse has already developed in this disease at the time of diagnosis, so treatment becomes difficult, and there is currently no uniform guideline for the treatment of this disease. The purpose of this review is to discuss the latest research progress of Kümmell's disease and provide some reference for spinal surgeons.

Keywords: Osteoporosis, Kümmell's Disease, Vertebral Fracture, Osteonecrosis, Intravertebral Vacuum Cleft

1. Introduction

Kümmell's disease (KD), a delayed complication of osteoporotic vertebral compression fracture (OVCF) [1], was first reported by Dr. Hermann Kummell in 1895 and is defined as individuals who suffer minor spinal trauma and are essentially asymptomatic for weeks to months, but then developed a symptomatic, progressive, angular kyphosis [2, 3]. There are also many names describing the disease, such as delayed post-traumatic vertebral osteonecrosis, intravertebral pseudarthrosis, intravertebral vacuum cleft (IVC), delayed vertebral collapse, and nonunion of compression fracture [4]. With the arrival of an aging society worldwide, the incidence of KD is increasing year by year. It has been documented that the incidence of nonunion is approximately 13.5% and that of intravertebral vacuum cleft (IVC) sign is about 7–13% in patients with OVCF [5]. At present, there are still controversies about the treatment options for this disease, and the following is a review of its clinical features, pathogenesis, radiological features, and treatment principles.

2. Clinical Features

Unlike OCVF, the most important clinical feature of KD is that after minor spinal trauma, most patients go through asymptomatic periods of weeks to months, followed by delayed vertebral collapse [6]. Only 23% to 33% of patients have clinical manifestations in the early stage, so it is difficult to diagnose early and fail to receive timely and effective treatment [7]. KD is common in women over 50 years of age, has a predilection for the thoracolumbar junction, and usually involves only a single vertebra, especially the T12 vertebra, and is the most commonly affected vertebral level [8]. The most commonly used clinical classification of KD is 5 stages of Steel [6] and 3 stages of Benedek [9].

In 1951, Steel classified KD into the following five stages, which are the initial insult (stage 1), post-traumatic period (stage 2), latent interval or stage of relative health state (stage 3), recrudescent stage (stage 4), and terminal stage (stage 5) [6] (Table 1).

Table 1. *Five clinical stages of KD, as proposed by Steel [6].*

Stage	Clinical feature
Stage 1: the initial insult stage	The initial trauma varies in severity and type, and the injury is usually hyperflexion of the spine. X-rays showed normal vertebrae.
Stage 2: post-traumatic period stage	Patients may complain of minor back pain and the pain is usually concentrated at the accidental site of injury without any post-traumatic sequelae and is able to work.
Stage 3: latent interval or stage of relative health state	Following the post-traumatic period and usually lasting weeks or months before the onset of progressive disability, the patient is not incapacitated.
Stage 4: recrudescent stage	Patient complained of back pain. The symptomatology is at first well localized with tenderness, pain and a feeling of insecurity at the site of the pathologic condition. A gibbous may be evident as loss of stature progresses. The symptoms tend to become more peripheral with root pain.
Stage 5: terminal stage	It is characterized by the formation of a permanent kyphus or may be made manifest by progressive pressure on roots or spina cord.

Thirty years later Benedek et al retrospectively diagnosed KD through 3 stages. The first stage is back pain after minor back trauma, which resolves after a few days to weeks. The second stage is asymptomatic intervals of months to years. The final stage is the recurrence of pain and the development of kyphotic deformity in the absence of significant additional trauma [9] (Table 2).

Table 2. *Three clinical stages of KD, as proposed by Benedek et al [9].*

Stage	Clinical feature
1	Back pain after minor back trauma, which resolves after a few days to weeks.
2	Asymptomatic intervals of months to years.
3	The recurrence of pain and the development of kyphotic deformity in the absence of significant additional trauma.

3. Pathogenesis

Since Dr. Hermann Kümmell first reported and proposed the underlying pathogenesis in 1895, that is, initial minor trauma to the vertebral body is not sufficient to cause fractures, but nutritional impairment of the vertebral body. This results in softening and resorption or atrophy of the adjacent vertebral surfaces and a progressive pressure atrophy deveops [10]. More than 100 years later, the pathogenesis of KD has not yet been elucidated. In 1978, Maldague et al [11] first proposed the intravertebral vacuum cleft (IVC sign) and suggested that KD is caused by ischemic osteonecrosis, while indicating that the IVC sign is a specific sign of local bone ischemia, and that the intravertebral vacuum cleft is filled with air from extracellular fluid (90% nitrogen) [12]. As in KD, most vertebral compression fractures occur at the thoracolumbar junction, which increases stress at this site due to greater thoracolumbar mobility. On the basis of osteoporosis, it is more likely to lead to nonunion due to its greater stress after trivial injury [13, 14]. At the same time, it easily leads to the failure of stable healing of the broken end of the fracture, and even pseudoarthrosis formation [15, 16].

Some scholars believe that the following risk factors may affect vertebral bone mineral density or vertebral blood supply, such as long-term alcoholism, bone metabolism diseases, long-term use of glucocorticoids, radiation therapy, hemoglobinopathy, vasculitis, pancreatitis, cirrhosis, Gaucher disease, diabetes, sarcoidosis, and Cushing's disease, which may be the cause of KD [8, 12, 17].

In conclusion, the prevailing view remains that avascular osteonecrosis is the main cause of IVC sign, as intravertebral vacuum clefts are thought to be associated with local bone ischemia associated with nonunion vertebral collapse, a sign that is unlikely to appear in neoplastic or inflammatory vertebral collapse. Therefore, in clinical practice, identification of IVC sign should prompt avascular osteonecrosis and a high suspicion of KD [11].

4. Radiological Features

Adamska et al suggest that only 23% to 33% of KD patients have early clinical manifestations [7]. Because most KD patients have no significant back pain symptoms in the early stages, and some scholars believe that Plain radiographs and computed tomography were taken at intervals between back trauma and vertebral collapse, and no evidence of fracture or bone destruction was found [18]. Therefore, this disease is difficult to diagnose early. The presence of the intravertebral vacuum cleft sign should be highly suggestive of KD and is a sign of vertebral avascular osteonecrosis [19], with a specificity of 99%, a sensitivity of 85%, and a positive predictive value of 91% [20].

4.1. Plain Radiographs

Standard radiographic examinations for K ü mmell 's disease include standing anteroposterior and lateral X-rays, and collapse of the anterior third of the vertebral body is usually more severe than posterior [21]. Radiographs showed that the intravertebral vacuum cleft was a transverse, linear to semilinear and radiopaque shadow, and the cleft was replaced by the accumulation of gas in the center of the vertebral body or adjacent to one of the endplates [22]. Standing flexion and extension lateral radiographs are often required for IVC sign, and intravertebral cleft is seen to be larger in the extension lateral position and smaller in the

standing flexion position, but patients are often unable to cooperate due to pain [5].

4.2. Computerized Tomography (CT)

Unlike X-rays, CT can more clearly show the findings of fractures, identify a heterogeneous and irregular bone fragments, and accurately show the affected areas of fractures [20]. Dynamic flexion-extension CT enables more sensitive detection of intravertebral vacuum cleft signs and spinal instability [5]. The shape of the IVC sign on CT is divided into three types, which are linear (IVC is evenly and continuously distributed in the vertebral body), triangular (IVC is triangular in the anterior half of the vertebral body), and irregular (IVC is heterogeneous or has several lines but no continuous distribution) [23].

4.3. Magnetic Resonance Imaging (MRI)

Li et al has proposed a three-stage classification for KD based on plain radiographs and MRI findings in 2004 [24]. These 3 stages were: Stage I: plain radiography showed complete or slight compression, and MRI showed signs of osteonecrosis. Stage II: Vertebral collapse with dynamic mobility but intact posterior vertebral wall. Stage III: Collapse of the posterior wall of the vertebral body causing spinal cord compression. Currently this staging system is widely used to describe the natural course of Kümmell 's disease and guide management policies [24].

MRI showed low signal intensity on T1 and T2 weighted images when air appeared in the intravertebral vacuum cleft. However, when the intravertebral vacuum cleft is filled with fluid, there will be low signal intensity on T1 weighted images, high signal intensity on T2 weighted images and fat-suppressed sequence (TIRM/STIR) [25], and some scholars believe that it is the sign of avascular necrosis [20]. MRI can also show a “double line” sign on T2 sagittal images characterized by a low signal line corresponding to a vacuum slit and surrounded by a high signal intensity corresponding to intraosseous edema [25].

4.4. Radionuclide Bone Image

Radionuclide bone scan was normal before back injury occurred, but radionuclide uptake increased 3 weeks after injury and before vertebral collapse. The first example of KD phenomenon documented by radionuclide bone scan is also considered and further supports avascular necrosis of the vertebral body [18]. Radionuclide bone scan is considered to be one of the sensitive tools for early diagnosis of osteonecrosis in KD, similar to osteonecrosis of the femoral head, and large studies in this area can be carried out in the future to confirm it.

5. Treatment

At present, there are different opinions on the treatment of the disease, and no unified consensus has been reached. For patients with KD, conservative treatment includes absolute bed rest, lower back brace, anti-osteoporosis treatment, and analgesic drugs, which can moderately reduce pain, but these treatment modalities do not seem to achieve good results as the course of the disease progresses [26].

Li et al [24] proposed to divide KD into three stages, followed by: the first stage: wedge deformation of the anterior vertebral body, loss of height less than 20 percent, no IVC sign and dynamic mobility. The second stage showed a loss of more than 20 percent of vertebral body height, accompanied by dynamic vertebral instability, but the cortical bone of the posterior wall of the vertebral body remained intact. For the above two stages of treatment, most scholars recommend minimally invasive treatment, of which Percutaneous vertebroplasty (PVP) and percutaneous kyphoplasty (PKP) are the most widely used. Baoliang Zhang et al. [27] concluded that both procedures were equally effective in neurologically intact KD and PKP was associated with a lower risk of cement leakage but a greater cost. Yajian Wang [28] stated that bone-filling containers mesh (BFC) is designed to control the dispersion of bone cement in the vertebral body through mesh containers, and BFC is most likely to be the best procedure for the treatment of KD based on a comprehensive assessment of the effectiveness of improving the patient's symptoms and the safety of adverse events.

The final stage is severe anterior vertebral wedging with posterior vertebral fractures and spinal cord compression [24]. The treatment modalities at this stage have become the most controversial topic. Because in the terminal phase of KD, the most important features are posterior vertebral fractures and spinal cord compression, which are the main causes of disability in patients, the purpose of treatment at this stage is to decompress and stabilize the spinal cord regardless of the mode of treatment. There are many treatment methods for the terminal stage, such as anterior direct nerve decompression and reconstruction, posterior short-segment internal fixation combined with vertebroplasty, posterior internal fixation combined with percutaneous vertebroplasty, posterior cement augmentation screw fixation, combined anterior and posterior surgery and vertebral osteotomyde compression.

In summary, when selecting which surgical method to use, individualized treatment should be done for each KD patient according to many factors such as the patient's age, degree of surgical tolerance, their own underlying diseases and economic conditions. And some scholars believe that the decision to perform anterior or posterior surgery should be based on the surgeon 's experience preference [29].

6. Conclusion

At present, Kümmell’s disease is no longer a rare disease, and as a spinal surgeon, how to diagnose this disease early has become increasingly important, and radionuclide bone scan can be recommended for patients suspected of KD, which can be confirmed by large studies in this area in the future. Minimally invasive treatment is recommended for patients with stage I and II Kümmell’s disease, and bone-filling containers mesh (BFC) may be the best treatment. For final stage patients, the choice of

surgical approach should be based on physician preference and the patient 's individual circumstances. At the same time, all patients with Kümmell's disease need to ensure basic anti-osteoporosis treatment. Finally, avoid risk factors or lifestyle that may affect bone mineral density or vertebral blood supply, thereby preventing this disease.

References

[1] Cheng H, Wang G, Li T, et al. Radiographic and clinical outcomes of surgical treatment of Kümmell's disease with thoracolumbar kyphosis: a minimal two-year follow-up. BMC musculoskeletal disorders, 2021, 22 (1): 1-761.

[2] Brower A C, Downey J E F. Kümmell disease: report of a case with serial radiographs. Radiology, 1981, 141 (2): 363-364.

[3] Swartz K, Fee D. Kümmell's disease: a case report and literature review. Spine (Philadelphia, Pa. 1976), 2008, 33 (5): E152-E155.

[4] Li H, Liang C, Chen Q. Kümmell's Disease, an Uncommon and Complicated Spinal Disorder: A Review. Journal of international medical research, 2012, 40 (2): 406-414.

[5] Lim J, Choi S, Youm J, et al. Posttraumatic Delayed Vertebral Collapse: Kummell's Disease. Journal of Korean Neurosurgical Society, 2018, 61 (1): 1-9.

[6] Steel H H. Kümmell's disease. The American Journal of Surgery, 1951, 81 (2): 161-167.

[7] Adamska O, Modzelewski K, Stolarczyk A, et al. Is Kummell's Disease a Misdiagnosed and/or an Underreported Complication of Osteoporotic Vertebral Compression Fractures? A Pattern of the Condition and Available Treatment Modalities. J Clin Med, 2021, 10 (12).

[8] Laredo J. Expert's comment concerning Grand Rounds case entitled "Kümmell's disease: delayed post-traumatic osteonecrosis of the vertebral body" (by R. Ma, R. Chow, F. H. Shen). European Spine Journal, 2010, 19 (7): 1071-1072.

[9] Benedek T G, Nicholas J J. Delayed traumatic vertebral body compression fracture; part II: pathologic features. Semin Arthritis Rheum, 1981, 10 (4): 271-277.

[10] Kümmell H. Ueber die traumatischen Erkrankungen der Wirbelsäule 1). DMW - Deutsche Medizinische Wochenschrift, 1895, 21 (11): 180-181.

[11] Maldague B E, Noel H M, Malghem J J. The Intravertebral Vacuum Cleft: A Sign of Ischemic Vertebral Collapse. Radiology, 1978, 129 (1): 23-29.

[12] Ford L T, Gilula L A, Murphy W A, et al. Analysis of gas in vacuum lumbar disc. AJR Am J Roentgenol, 1977, 128 (6): 1056-1057.

[13] Kong L D, Wang P, Wang L F, et al. Comparison of vertebroplasty and kyphoplasty in the treatment of osteoporotic vertebral compression fractures with intravertebral clefts. Eur J Orthop Surg Traumatol, 2014, 24 Suppl 1: S201-S208.

[14] Li H, Liang C, Shen C, et al. Decreases in fluid shear stress due to microcracks: A possible primary pathogenesis of Kümmell's disease. Medical hypotheses, 2011, 77 (5): 897-899.

[15] Matzaroglou C, Georgiou C S, Assimakopoulos K, et al. Kümmell' s disease: A rare spine entity in a young adult. Hellenic journal of nuclear medicine, 2010, 13 (1): 52.

[16] Ranjan M, Mahadevan A, Prasad C, et al. Kummell's disease - uncommon or underreported disease: A clinicopathological account of a case and review of literature. J Neurosci Rural Pract, 2013, 4 (4): 439-442.

[17] Matzaroglou C, Georgiou C S, Assimakopoulos K, et al. Kümmell's disease: pathophysiology, diagnosis, treatment and the role of nuclear medicine. Rationale according to our experience. Hellenic journal of nuclear medicine, 2011, 14 (3): 291.

[18] VAN EENENAAM D P, EL-KHOURY G Y. Delayed post-traumatic vertebral collapse (Kummell's disease): case report with serial radiographs, computed tomographic scans, and bone scans. Spine (Philadelphia, Pa. 1976), 1993, 18 (9): 1236-1241.

[19] Theodorou D J. The intravertebral vacuum cleft sign. Radiology, 2001, 221 (3): 787-788.

[20] Libicher M, Appelt A, Berger I, et al. The intravertebral vacuum phenomen as specific sign of osteonecrosis in vertebral compression fractures: results from a radiological and histological study. Eur Radiol, 2007, 17 (9): 2248-2252.

[21] CHOU L H, KNIGHT R Q. Idiopathic avascular necrosis of a vertebral body: Case report and literature review. Spine (Philadelphia, Pa. 1976), 1997, 22 (16): 1928-1932.

[22] Yu C W, Hsu C Y, Shih T T, et al. Vertebral osteonecrosis: MR imaging findings and related changes on adjacent levels. AJNR Am J Neuroradiol, 2007, 28 (1): 42-47.

[23] Chen Z, Lou C, Yu W, et al. Comparison of Intravertebral Clefts between Kummell Disease and Acute Osteoporotic Vertebral Compression Fracture: A Radiological Study. Orthop Surg, 2021, 13 (7): 1979-1986.

[24] Li K, Wong T, Kung F, et al. STAGING OF KÜMMELL'S DISEASE. Journal of Musculoskeletal Research, 2011, 08 (01): 43-55.

[25] Nickell L T, Schucany W G, Opatowsky M J. Kummell Disease. Baylor University Medical Center Proceedings, 2017, 26 (3): 300-301.

[26] Yoon S T, Qureshi A A, Heller J G, et al. Kyphoplasty for salvage of a failed vertebroplasty in osteoporotic vertebral compression fractures: case report and surgical technique. J Spinal Disord Tech, 2005, 18 Suppl: S129-S134.

[27] Zhang B, Chen G, Yang X, et al. Percutaneous Kyphoplasty Versus Percutaneous Vertebroplasty for Neurologically Intact Osteoporotic Kümmell's Disease: A Systematic Review and Meta-Analysis. Global Spine Journal, 2022, 12 (2): 308-322.

[28] Wang Y, Liu B, Sun Z, et al. Comparative Efficacy of Three Minimally Invasive Procedures for Kümmell's Disease: A Systematic Review and Network Meta-Analysis. Frontiers in surgery, 2022, 9: 893404.

[29] Pappou I P, Papadopoulos E C, Swanson A N, et al. Osteoporotic vertebral fractures and collapse with intravertebral vacuum sign (Kummel's disease). Orthopedics, 2008, 31 (1): 61-66.