Kümmell' s disease: A rare spine entity in a young adult

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Abstract

Over 100 years ago, Dr Hermann Kümmell described a rare clinical entity in which patients, after a trivial trauma and an asymptomatic period, developed a progressive vertebral body collapse and a painful kyphosis. *We present the case* of a 31years old heavy labourer, fitting Kümmell's criteria. The patient referred to us in an incapacitated state, due to persistent back pain. Radiographic examination revealed a body collapse of L1 vertebra. The patient had no previous medical record, other than a prolonged history of transient back pain episodes, related to heavy-weight lifting. Last attack was 1 year before presentation. Through course of time, he had undergone *several clinical and radiological evaluations*, by different orthopaedists, on different occasions, including the last episode, with no major findings. After an extensive workup, a percutaneous kyphoplasty of the affected vertebra was performed and a biopsy was obtained. The histologic examination of the specimen revealed vertebral osteonecrosis. A triggering pattern of repetitive spinal loading in hyperflexion is, for the first time, being recognized. *We conclude* that Kümmell's disease, although a rare condition, should be considered in any patient with refractory back pain symptoms. In such patients, vigorous follow-up turns to be of the essence.

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Introduction

Before the advent of X-rays, Dr Hermann Kümmell in 1891 described a series of five patients, who developed a posttraumatic vertebral collapse, after a trivial spinal trauma and after an essential asymptomatic period of months to years. For a period of 35 years, after the original dissertation, Kümmell's disease (KD) was struggling for acceptance [1]. It was only with the advent of radiographic technology that this concept was validated. Thereafter, this eponymous diagnosis represented posttraumatic vertebral fractures, which were initially asymptomatic, with unremarkable initial X-rays findings, and by later with vertebral body collapse. In recent years, KD was defined as delayed posttraumatic vertebral body collapse, because of showing avascular osteonecrosis, which predominantly affected the lower thoracic and the upper lumbar vertebrae [2, 3]. Originally, KD involves patients, without any other evident cause for ischaemic necrosis, other than trauma. In the past 50 years, there have been at most 10 cases reported, meeting Kümmell's criteria [2].

We present a rare case of a 31 years old male, heavy labourer fitting the criteria of KD. To our knowledge, our case is the first description of KD in a relatively young individual, after Steel's report (1951) in a 23 years old patient [1]. Repetitive heavy-weight lifting and thus, hyperflexion as a spinal strain, is for the first time recognized as a pattern of spinal injury, that could eventually cause KD.

Description of the case

A 31 years old male construction worker referred to our special Spinal Surgery Unit with an incapacitated back pain, after involved repetitive heavy-weight lifting. The patient had a clear medical record and was under no medication. He was afebrile, mentioned no body-weight loss in the last semester but described a prolonged history of back pain episodes, during the last 5 years. Every episode started after lifting a heavy object and the pain settled with analgesics. For some of these attacks, the patient was examined by different orthopaedists and had undergone several plain radiograms of the spine -which the patient never stored- in various instances with no major findings. His symptoms were attributed to muscle contraction, based on the absence of radiculitis, patient's young age and fast response to analgesics. No computed tomography (CT) or bone scan was ever obtained. This was also

the case for the last episode, a year ago. The mechanism was also the same; heavy-weight lifting with hyperflexion of the spine. The patient had persistent back pain for two days and then visited a private orthopaedist. Radiographic examination was again without any findings. The symptoms however, never really resolved. The inconsistent mild mid back pain at first, worsened progressively in intensity and frequency. During the last month the back pain started limiting his ability to walk and was localized to the area of the thoracolumbar junction with no pain radiation or muscle weakness.

On admission, laboratory tests showed a leukocyte count of 5.93k/μL (normal, 4-11k/μL), erythrocyte sedimentation rate of 23mm Hg, on the first hour (normal, 1-35mm/hr), C-reactive protein: 3mg/L (normal, 1-10mg/L), normal tumor markers (CEA, aFP, βhCG, PSA, CA 19-9), normal serum electrophoresis and normal bone (alkaline phosphatase, serum Ca and P) and liver tests (transaminases, total and direct bilirubin, albumin). Chest radiogram was also normal. A skin test for tuberculosis was negative. Anterior posterior (AP) and lateral plain radiographs of the thoracolumbar junction were obtained. The AP radiograph showed collapse of the body of the L1 vertebra and intact pedicles. The lateral radiograph showed anterior wedging of L1 vertebral body, with no retropulsion of the posterior vertebral margin (Fig. 1). A CT and a magnetic resonance imaging (MRI) scan of the thoracolumbar spine followed (Fig. 2). Both showed a destructive lesion of the anterosuperior part of the L1 vertebral body with no involvement of the pedicles, of the posterior elements or the paravertebral soft tissues. The lesion showed no contrast enhancement on CT images (Fig. 2A), while on MRI scan low signal intensity on both T1 and T2-weighted sequences (Fig. 2B). A technetium-99m methylene diphosphonate (^{99m}Tc-MDP) bone scan was, also, performed 3h after the intravenous injection of 740MBq and demonstrated a barely visible increase of uptake in the selected region of interest (ROI). Subsequently, a single photon emission tomography/CT (SPET/CT) study was performed, which did not essentially enhance on the planar bone uptake imaging, although the fused SPET/CT images (Fig. 3) confirmed the findings of planar imaging and provided the exact, 3D, lesion position. The meagre uptake of the osteophilic radiopharmaceutical was consistent with the chronicity of the lesion, as the normal osteoblastic response within the osseous reconstruction phase would have been indicative of a recent lesion. Additionally, the bone scan did not reveal any other sites of selective focal uptake, thus, diminishing the possibility of the lesion being of a secondary (metastatic) nature. During a psychiatric interview, health related quality of life (HRQoL) was assessed using the Greek version of the short form-36 health survey (SF-36), which is a self-report measure, covering eight domains of HRQoL, that evaluate the extent to which an individual's health limits his or her physical, emotional, and social well-being [4]. The Greek validated version of the hospital anxiety and depression scale (HADS) was also completed [5]. This questionnaire assessed both depression and anxiety. Analysis of results showed that the patient is HRQoL were severely affected. He was, also, diagnosed as having moderate anxiety and depression.

After this extensive workup, surgical intervention was decided. A percutaneous kyphoplasty of the L1 vertebra was performed (Fig. 4) and a biopsy was obtained. Histologic examination revealed necrotic bony trabeculae and no signs of neoplasia or infection. Aerobic, anaerobic, acid fast bacilli and fungal cultures of the specimen showed no growth. Polymerase chain reaction for tuberculosis was also negative. The diagnosis of KD was established.

Patient's pain gradually diminished and eventually disappeared three months postoperatively. The patient was followed-up with a psychiatric examination as previously described at one and six months post-kyphoplasty. His HRQoL returned to normal, whereas no evidence of depression or anxiety remained.

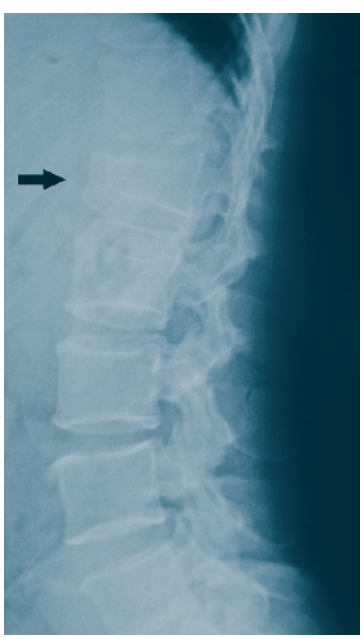
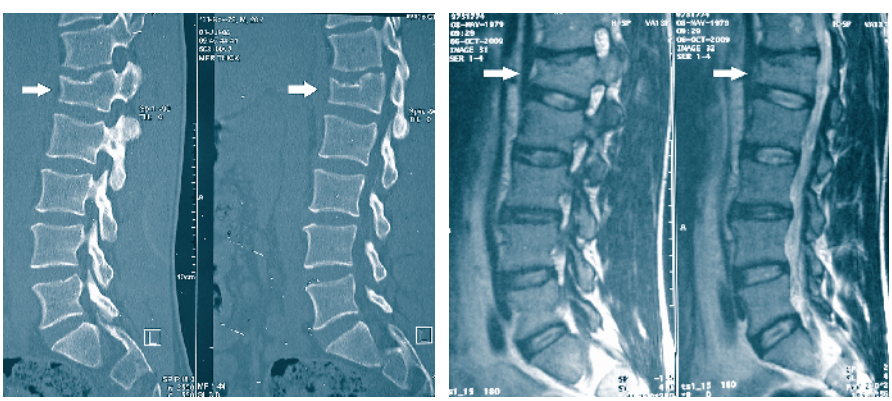


Figure 1. Plain lateral radiograph shows, anterior wedging of the L1 vertebral body, with no retropulsion of its posterior vertebral margin.



A B

Figure 2. Computerized tomography (A) and MRI (B) scans of the thoracolumbar spine. A destructive lesion of the anterosuperior part of the L1 vertebral body is depicted. (A): No contrast enhancement of the lesion is seen on the CT scan. (B): Low signal intensity on the T2-weighted sequences of the MRI images.

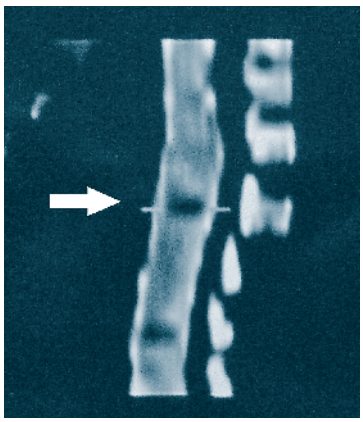


Figure 3. Meagre uptake of the osteophilic radiopharmaceutical in the SPET/CT bone study, is seen, which is consistent with the chronicity of the lesion.

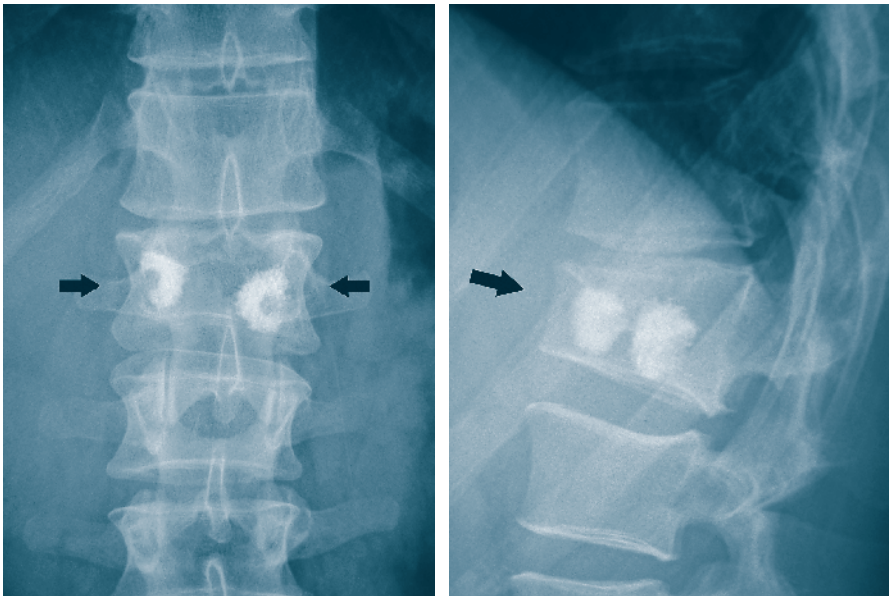


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Figure 4. Postoperative plain radiographs of the thoracolumbar spine. Anterior-posterior (A) and lateral (B) views. Notice the restoration of the anatomical shape of L1 vertebra.

Discussion

In 1891 a German surgeon, Dr Hermann Kümmell described a series of 5 patients, presenting a rare clinical entity: they sustained a minor spinal trauma, then remained essentially asymptomatic for a period of months or years and eventually, developed a progressive, painful kyphosis at the lower thoracic or upper lumbar regions [1]. With the advent of X-rays, it was recognized, that the angular deformity was the result of a delayed vertebral body collapse (VBC). It was not until 1951, that Steel further divided the clinical course of KD into five stages [1]. The initial injury can be varied in severity, while lateral roentgenograms are necessarily negative. Post-traumatic period follows with minor complaints and no limitation in activity. The latent interval or third stage of relative well-being, usually lasting weeks or months, during which time the patient is not incapacitated, follows the post-traumatic period and precedes the onset of progressive disability,. In the fourth stage, the recrudescent stage, the patient complains of persistent, localized pain, which progressively tends to become more peripheral and with root pain. In the last stage, the terminal stage, a permanent kyphus is formed and/or spinal cord compression is established. Kümmell's disease occurs typically in middle-aged and elderly patients with a slight male predominance [1]. The only exception is Steel's report on a 23 years old patient in 1951 [1]. Since then, our case is the first description of the disease in a relatively young patient.

Although more than a century has passed, since Kümmell's disease was described, there is still no unanimous consensus for the pathogenesis of this entity. Early hypothesis described multiple, minute traumas of osseous and ligamentous structures resulting in fine cracks and microhemorrhages; these minute ruptures in the spongiosa lead to osteonecrosis. The VBC is the result of too early strain on deficient material [1]. Benedek and Nicholas (1981) suggested that the initial trauma caused trabecular fractures in the vertebrae, which were unremarkable on radiograph analysis; they hypothesized that a second phase occurred, characterized by impaired healing, because of vascular supply disruption. Because of impaired healing, the trabecular fractures developed into a VBC [6]. Another hypothesis, based on spinal angiography findings, attributes VBC to vascular insufficiency. The anterior third of the vertebral body (VB) is described as a vascular border zone/"watershed". The dorsum of a VB receives collateral blood flow, while the ventral aspect does not, putting the latter at higher risk for ischaemic necrosis [2, 7]. Others on the basis of pathologic analysis of specimens from patients with delayed posttraumatic VBC, proposed the concept of osteonecrosis after minute ruptures and hemorrhage into the spongiosa [8].

Currently, avascular osteonecrosis is the predominant hypothesis for the interpretation of the delayed posttraumatic VBC [3]. Injury seems to trigger vascular supply disruption in KD [1]. Trauma, however, can be of different natures. Originally Kümmell, in his series, has described a hypeflexion pattern of spinal injury, after a fall. Others reported a case with symptoms onset related to lifting a heavy object [9] or another case without any predisposing factors and with symptoms triggered after shoveling snow [3]. These two reports attribute VBC not to direct or indirect trauma, but to hyperflexion loading of the spine. However, they differ from our report because they indicate that heavy-weight lifting was incidental. In our case repetitive weight-lifting and thus hyperflexion spinal strain was the causative pattern for KD. This mechanism is for the first time described.

As far as diagnosis of KD is concerned, it remains a diag-

nosis of exclusion. There is no pathognomonic radiographic finding. The best testing is serial imaging, depicting an initially intact VB after trauma, and then VBC, as the ischaemic necrosis develops. Another radiographic finding, an intravertebral vacuum cleft on plain radiograph and CT scan, has been associated with avascular osteonecrosis [10]. However, this finding can also be seen in other conditions [11] or simply reflect gas migrating from an adjacent affected disc into the VB [12]. On MRI, intravertebral vacuum cleft has an increased signal on T-1 weighted images, and a decreased signal on T-2 weighted images; occasionally a peripheral zone of hypointensity can be identified around the hyperintensity on T-2 weighted images [13]. This hyperintensity with surrounding band of hypointensity on T-2 weighted images is termed "double line sign" and represents sclerosis surrounding central granulation tissue [2]. However, in patients in later phases of avascular necrosis of the VB, like our patient (Fig. 2A), a linear area of hyperintensity on T2-weighted images could not be identified [13].

Once VBC has been demonstrated, a thorough history and general medical evaluation must be obtained. Because VBC can be seen in a variety of conditions including neoplasms, infections and osteoporosis, testing should include: complete blood count, complete metabolic panel, erythrocyte sedimentation rate, and a MRI with and without contrast. Depending on the findings, other testing, such as bone scans and biopsy may be indicated. Bone scans and especially four phases bone scans have been shown to demonstrate abnormalities during the early phase of KD, when plain X-rays are normal [9]. Comparison with old films may help establish whether a compression fracture is acute or chronic, but in the absence of relevant films, a bone scan or a MR image can help. Bone scan will show absent or minimal uptake if the lesion is chronic [14].

Treatment is dependent on patient's symptoms and test findings. For acute VBC with back pain and with the prerequisite of an intact posterior VB wall, kyphoplasty or vertebroplasty can be considered. For chronic VBC or acute VBC in which the posterior wall is disrupted, surgical stabilization via fusion is indicated. If there is evidence for neurologic compromise, then decompression with stabilization should be performed expeditiously [2].

In conlusion, KD is a delayed post-traumatic vertebral body collapse. Our case indicates that although KD is a rare condition, it should be considered in every patient, with refractory back pain problems, especially when having a history of spinal trauma or prolonged hyperflexion loading. These patients should be regularly reevaluated radiographically or by scintigraphy. If a VBC is recognized, patients should undergo an extensive work-up to exclude other potential underlying conditions, before a definite mode of treatment is decided.

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