Commentary

In 1891, Dr. Hermann Kümmell, a German surgeon, described a clinical entity characterized by the development of progressive painful kyphosis following an asymptomatic period of months or years after a minor spinal trauma. With the advent of radiography in the last century became evident that cause of the kyphosis described by Kümmell was a delayed vertebral body collapse.[1] This collapse represents a failure of the fracture healing process due to the development of an avascular necrosis zone below the superior endplate, resulting in an atrophic or avascular non-union, so this disorder is currently known as Kümmell's disease (KD) or vertebral ostenecrosis.[2]

KD usually occurs in adults older than 50 years of age, mostly women, and involves more frequently only one vertebral body at the T8 through to the L4 level, with the anterior third of the vertebral body being the most affected.[3] Although the patients remain asymptomatic or with mild pain for a variable period of time, they experience worsening of pain when collapse develops, that might be severe and limiting, and associate neurological complications.[2-4] KD is not commonly associated with classic risk factors for peripheral osteonecrosis such as steroid therapy or alcoholism, but it more frequently occurs in patients with history of vertebral compression fractures.[5]

The diagnosis of KD is difficult because there are no pathognomonic radiographic findings, so it is considered as a diagnosis of exclusion.[6] A characteristic radiological sign of KD is the so-called "vacuum cleft sign," an intravertebral air collection recognizable as linear or semi-lunar radiolucent shadow on plain X-rays, or with a more heterogeneous distribution and an irregular shape in computed tomography scans. This sign is highly suggestive of KD, having been demonstrated as a strong correlation with biopsy-proven osteonecrosis, and a high sensitivity and specificity. However, this sign is often difficult to identify and might appear only on hyperextension radiographs; therefore, its absence does not exclude KD diagnosis.[3] Furthermore, this sign might also be seen in other conditions such as malignancy or intraosseous disc prolapse (Schmorl's nodes).[4] The best testing is serial imaging, showing an initially intact vertebral body after trauma, and then its collapse when the avascular necrosis develops.[6] Magnetic resonance might also contribute to the diagnosis of KD. The vacuum cleft is observed as a zone with low-signal intensity in all sequences, or might secondarily be filled with fluid and appear as a well-circumscribed area of low-signal intensity on T1-weighted and high signal intensity on T2-weighted images. This finding is called the "fluid sign."[3]

KD has been long considered a rare entity because since its original description has been reported sporadically in the literature, mainly as short series or case reports. This fact has been reinforced due to multiple terms used to describe it, including avascular necrosis of the vertebral body, vertebral pseudarthrosis, intervertebral vacuum cleft or gas, delayed vertebral collapse, and vertebral compression fracture non-union. However, KD is actually underdiagnosed, and its true incidence is quite high, representing 7% to 37% of all vertebral compression fractures.[2]

The knowledge of KD is important because is associated with neurological complications more often than osteoporotic compression fractures, so the delay in diagnosis may lead to the development of serious sequelae.[2-4] KD should also be considered in the differential diagnosis of spinal infections.[4] Ranjan *et al*,[7] presents a very interesting case that pose a diagnostic dilemma with tuberculosis in an endemic environment. Spinal tuberculosis accounts up to 59% of all orthopedic

tuberculosis, involving more frequently the vertebral body and following a progressive course. The destruction of the vertebral body is secondary to the infection, but avascular necrosis is also involved.[8] KD and spinal tuberculosis share some clinical features such as kyphosis development and neurological complications, and spinal tuberculosis also involves some segments where KD occurs, being L1 the most commonly affected body and T10 the most commonly associated with cord compression.[3,4,8] While in the spinal tuberculosis, the cornerstone of treatment is drug therapy and the anterior fusion operation,[8] there are no specific guidelines for KD treatment, and current management follows an approach similar to osteoporotic compression fractures, which is to be initially conservative. However, recent reports favor surgical interventions for prevention of neurological complications. Minimally invasive procedures, such as vertebroplasty, have also been used for pain relief in refractory cases, with similar results to osteoporotic fractures.[3]

In conclusion, KD is not rare and should be considered in the differential diagnosis of vertebral compression fractures and spinal tuberculosis, allowing keep close monitoring for prevent neurological complications in the first, and to avoid delay in diagnosis and unnecessary treatments and interventions in the second.

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