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Kummell's Disease, Case Report of a Delayed Onset Paraparesis and Literature Review

Salvatore D'Oria*, Carlo Delvecchio, Francesco Zizza, Carlo Somma
Neurosurgical Unit of Miulli Hospital of Acquaviva delle Fonti, Bari, Italy

Abstract

Introduction: Kummell's disease is an avascular necrosis of the vertebral body, secondary to a vertebral compression fracture. This entity is characterized by the gradual development in time of a vertebral body collapse following a trivial spinal trauma, involving a worsening back pain associated to a progressive kyphosis. Following the progressively ageing population the prevalence of osteoporosis is increasing and as a consequence, the incidence of spinal crush fractures; therefore evidence of Kummell's disease is quite common, also favoured by the great accuracy of modern diagnostic equipment.

Purposes: The aim of this article is to carry out an international literature review regarding Kummell's disease, addressing its physiopathology, histopathology, clinical presentation, radiological characteristics and treatment modalities; at the same time, literature is updated through the description of a new and interesting case, symbol of the pathology long- term potential complications, if not diagnosed and therefore not suitably treated.

Case report: A patient with osteoporosis, following a slight spinal trauma, suffered a progressive necrosis of the D11 body; although the radiological exams showed a constant worsening of the thoracic lumbar kyphosis and a restriction of the spinal canal, in another medical centre he was only treated with a corset and painkillers. A year after the injury, motor deficits concerning the lower limbs appeared. He was then sent to us and indication for posterior internal fixation was given. On the basis of both his medical history and radiological and histological findings, Kummell's disease was diagnosed.

Conclusion: Is necessary to have a complete knowledge of the clinical, pathological and radiological characteristics of Kummell's disease, as to follow a correct diagnostic course enabling to prepare the most suitable therapy.

Keywords

Kummell's disease; Progressive kyphosis; Spinal trauma; Histopathology; Physiopathology

Abbreviations: KD: Kummell Disease; AVN: Avascular Necrosis; VFC: Vertebral Compression Fracture; IVC: Intravertebral Vacuum Cleft; VBC: Vertebral Body Collapse; CT: Computer Tomography; MRI: Magnetic Resonance Imaging

Introduction

Kummel's disease (KD) or avascular necrosis (AVN) of vertebral body was described for the first time by the German surgeon Hermann Kummell, who defined it as a vertebral body collapse after a minor vertebral trauma taking place long after the traumatic event, thus becoming symptomatic only after a certain amount of time [1,2]. Even if more than a century has elapsed from the first description of this pathology, currently there still are only few reports dealing with this matter; in fact, according to a previous review, ten years ago, only 120 articles were available on this subject [3]. It is however believed that KD prevalence is higher than what appears in literature, and it is also underestimated for the lack of a sufficient knowledge of the same by doctors, as well as for an heterogeneous terminology referred to it: vertebral compression fracture (VCF) nonunion, intervertebral vacuum cleft, delayed vertebral collapse, post-traumatic vertebral osteonecrosis, *avascular necrosis* and vertebral pseudoartrosis [4]. On the other hand, it must be remarked that in the past many authors mistakenly presented KD cases in which vertebral necrosis was due to other diseases and not correlated to traumas [5]; in this regard, an element of confusion is constituted by the intervertebral vacuum cleft (IVC) which is frequent in KD, but not pathognomonic for it, as it can be found, albeit rarely, in other types of necrosis [6]. Furthermore there is disagreement on the correct eponymous, as various authors adopted the terms Kummell, Kummel or Kummell-Verneuil [7]. The most affected age group is the medium-high one, in which often osteoporosis coexists, representing a risk factor for VCFs and therefore for AVN; so, following the progressive ageing population, today AVN prevalence is increasing, amounting to a percentage varying between 7% and 37% [8,9]; between the 2 genders, it has been registered a slightly higher incidence in men [10]. Besides osteoporosis, other risk factors include chronic therapy with steroids, alcoholism and pre-existing radiotherapy [3]. The most credited pathogenic hypothesis today, as it was originally guessed by Kummell, consists in a loss of the vertebral vascular support, following low energy spinal injuries, leading to osteonecrosis and compromising the break healing process [4,7]. In this report we analyse KD clinical and radiological aspects and we show the relative therapeutic options trying to put some order among the concepts, sometimes dissenting, that are present in literature. Moreover we present a case of a patient who encountered paraparesis precisely because he had not received a right diagnosis and he was therefore treated in an unseemly manner.

***Corresponding author:** Salvatore D'Oria, Department of Neurosurgery, Second University of Naples, SUN, Italy, Tel: +3299282013; E-mail: doriantonioraf@libero.it

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Method

This paper drew on from the articles dealing with KD present on Pubmed, Medline and Google Scholar; the following key words have been used: Kummell's disease, vertebral avascular osteonecrosis, intravertebral vacuum cleft, review, case report. The necessary data for the drafting of this article have been extracted and summarised from the retrieved scientific material.

Background

The German surgeon Hermann Kummell (1852-1937) described for the first time the nosographic entity under discussion in 1895. Its case history included six patients who, after having undergone a slight vertebral trauma, complained, after a first phase, of spinal pain, however they then remained asymptomatic over months or years until a second phase characterized by pain relapse; during the third and last phase a kyphosis developed [1,2]. In 1911, one of Kummell's students assigned the name of his teacher to this pathology. Some years before the French surgeon AA Verneuil (1823-1895) had illustrated a similar clinical condition, therefore sometimes we speak about *Kummell-Verneuil's disease* [11].

In a later period it became possible to study the vertebral column with x-rays that showed how in the first phase of this pathology vertebral lesions weren't evident while, later on, there was a vertebral body collapse (VBC) [10,12].

More than 100 years have elapsed after the determination of this pathological process, however, if we analyse scientific literature, it is possible to come across only a few cases that respect clinical, radiological and histological criteria to diagnose KD, in particular it is necessary the occurrence of a spinal trauma with initial negative radiological study and only after, VBC with osteonecrosis [13]: Young [7], analysing English literature from 1950 until the sunset of the twentieth century, found only 5 cases, while in a review by Matzaroglou in 2014 [13] only 9 were identified.

Aetiopathogenesis and Risk Factors

- Risk factors are represented by all those conditions making the vertebra frail, such as osteoporosis, chronic use of steroids, cancer and radiotherapy.
- A vertebral trauma determines the reduction of the blood contribution to the bone with the following necrosis: the scarcity of the cancellate bone trabeculae leads to a vertebral body collapse. Restorative bone processes of the collapsed vertebra follow, which can lead to an excess osteogenic reaction provoking a restriction of the vertebral canal and therefore compression of the nervous structures.
- Several pathogenetic hypothesis have been proposed regarding the mechanisms underlying KD. Kummell suggested that the loss of nutritional contribution of the bone would have caused fragility and bone reabsorption leading, in a second moment, to a vertebral body collapse [1]. Atrophic nonunion [4], microfracture [11], fatigue fracture [14], pseudoarthrosis [15] and avascular necrosis [16,17] were then brought into play. Especially this last theory is highly prevalent today, supported by evidence, during histology, of necrotic bone fragments immersed in a stroma of fibrous reactive tissue [5,7,18,19]. Necrosis would be secondary to the vascular contribution impairment [20]; an angiography study showed in three patients, occlusive phenomena and rarefaction at the expense of arterioles in the portion of the collapsed vertebra [21], furthermore the vertebral body collapse usually takes place in the soma anterior third, which, constituting a watershed zone, is particularly sensitive to ischemic strokes [16]. The correlation between KD and bone necrosis is furthermore supported by the coexistence, noticed in three patients, of vertebral and femoral head necrosis [22,23] as well as by the radiological similarities among crescent signs, found in avascular necrosis of the femoral head and IVC, typical of KD [23]; it must be remembered that in case of vertebral osteonecrosis, a gas collection is often set up which on the contrary is not associated to femur necrosis [23].

Whereby AVN takes place, it is necessary that a traumatic event acts on the vertebral body [7], usually with a hyper flexion mechanism, in a predisposed individual; in fact, if a vertebral necrosis with a negative medical history for trauma takes place, by definition we cannot speak about KD. The reparative bone processes of the collapsed vertebra can lead to an excess osteogenic reaction provoking a restriction of the vertebral canal and therefore compression of the nervous structures.

The main risk factor for vertebral body AVN is osteoporosis [4] which determines rarefaction of spongy bone trabecular microarchitecture, making it brittle and therefore favouring, after trivial traumas, hairline fractures, responsible for injuries of intramedullary arterioles [11]. As KD usually develops in osteoporotic individuals, it is rare its occurring in a young subject, there is only one documented case in which it was diagnosed to a young male [24]. Another predisposing factor is constituted by the chronic use of steroids which would favour the intramedullary fat deposition compressing and obstructing the vessels [25,26]; other morbid conditions associated to KD include the sickle trait [8], as haemoglobinopathies favour vessel obstruction, pancreatitis, Gaucher's disease [27,28], sarcoidosis [29] and in general all factors that make the vertebra fragile as cancer and radiotherapy or that facilitate vessel obstructions as atherosclerosis, diabetes mellitus and vasculitis [16,30].

Clinical Features

- Kummell's disease affects advanced age people with a slight predominance of the male gender and it is generally located in the thoracic lumbar tract, normally concerning one single vertebra.
- The typical course of the disease foresees a first phase corresponding to the traumatic event, which can be more or less painful; the painful symptoms slowly fade away followed by a period of relative wellbeing. After few months, resurgent spinal pain (and sometimes radicular) appears, finally neurological deficits and vertebral deformities arise.

In 1951 Steel subdivided the course of the pathology into 5 phases [10]

I) Initial injury: variable intensity spinal pain but negative radiograms.

II) Post traumatic period: minor nature symptoms, without any disability.

III) Latent interval: relative wellbeing having a variable duration equal to few weeks or several months.

IV) Stage of resurgence: appearance of persistent back pain, sometimes associated to radicular pain.

V) Terminal stage: spine kyphosis sometimes followed by compression of the nervous structures.

Kummell's disease affects advanced age people with a slight predominance of the male gender [10]. There is only one documented case in which it was diagnosed to a young male [24]. The mostly affected area is the thoracic lumbar one; in fact, representing a transition segment between the thoracic tract, which is stiff, and the lumbar, which is mobile, it is particularly liable to traumatic strengths [30]. Generally only one vertebra is affected, and this vertebra is included between T8 and L4, but most frequently (60%) it is found between T11 and L1 [31], maximum at T12 [32]; in rare cases multiple spinal metamers are

involved [5,8]. The characteristic clinical course is connoted by a severe spinal pain, however having a moderate intensity, usually perceived at a lower thoracic level or upper lumbar, following a slight trauma, commonly collateral to a fall, without any radiological alterations. This is followed by an asymptomatic period, which generally lasts for 13 months, up to 8 years [16]. Finally the pain reoccurs, this time without any correlation to a new trauma, and generally with an intensity greater than the previous episode, and there is the development of a kyphosis with its core corresponding to the collapsed vertebra; at the same time the vertebral canal can shrink, determining neurological deficits as for example hyposthenia and paraesthesia to the lower limbs and intestinal or bladder disorders.

Radiologic Aspects

- In the initial stages of the disease, no pathological specimens arise from the radiological exams, except potential small fractures detectable through computer tomography.
- Once the asymptomatic period has elapsed, with the resurgence of pain, the x-ray shows a vertebral somatic collapse and the vacuum cleft as a transverse radio opaque area in the centre or at the periphery of the soma, indicating the entrance of gas in the vertebra. During the CT the vacuum cleft is not linear but it presents an irregular morphology. The "fluid sign", namely the cleft filled with fluid (hyper intense in T2) is typical of RM, because of the patient's supine position and sometimes the "double line sign", constituted aforementioned fluid sign surrounded by an hypo intense halo, to be referred to sclerosis.
- During the advanced phase of the pathology, neuro-radiological inquiries will stress spine kyphotisation, deformity following bone reparation phenomena of the involved vertebral body and reduction of the spinal canal diameters.
- It must be stressed that the vacuum cleft is not pathognomonic of Kummell's disease; instead it constitutes a mere osteonecrosis marker.

In accordance with the trivial nature of the initial trauma, the x-ray carried out immediately after the same is usually negative, however a computer tomography (TC) acquired during the early phases of the disease could reveal some initial bone alterations as for example small fractures [25]. Instead they performed x-ray examination, usually taking place in connection with back pain resurgence, shows a crush of the involved vertebral body, at the anterior portion of the same or anyway mainly anterior [16,23]. At this point the x-ray would show the typical vacuum cleft, marker of vertebral osteonecrosis [23,33] and due to the entrance of gas in the vertebra, as a transverse and linear radiolucency in the middle of the soma or adjacent to one of the endplates [23]; the IVC is filled for its 95% with nitrogen and peat, the remaining part with oxygen and carbon dioxide [34]; in dynamic x-rays the IVC appears mobile shrinking in bending and widening in extension, simultaneously the vertebral body is reduced or grows in height [35]; sometimes in bending the IVC disappears [23]; the above mentioned phenomenon indicates instability of the fracture [36] and a greater probability of developing chronic pain [6].

During the CT, the IVC presents a different morphology compared to the x-ray, characterized by widespread distribution and irregular shape.

Magnetic resonance (MRI) shows the necrotic region as an area with a signal increase T1-weighted images and of decreased signal on T2-weighted images, on the contrary, the simple chronic osteoporotic collapse does not determine signal changes on MRI [16,37]; the IVC is represented by a strip of hypo intensity in both T1 and T2 sequences, however, during images acquisition, due to the patient's supine position, the cleft is filled with fluid, which appears hyper intense in T2 [38,39], this is the fluid sign [40]; sometimes the hyper intense area in T2 is surrounded by a hypo intense halo corresponding most likely to sclerosis, in fact, biopsies of this area clarified its nature of fibrocartilage immersed in fibrous stroma [41], such fins is defined double line sign [39] and it can be found in the femoral head necrosis [5,42].

During the advanced phase of the pathology, neuro-radiological inquiries will stress spine kyphotisation, deformity following bone reparation phenomena of the involved vertebral body and reduction of the spinal canal diameters.

It must be underlined that there aren't any radiological frameworks pathognomonic of KD, in fact the IVC, despite being strictly correlated to it, it does not have its exclusivity, simply indicating a vertebral osteonecrosis [39] where the ischemic collapse of the bone substance leads to the cleft formation, which presents low pressure and therefore brings gas within itself [43]. Libicher [44], analysing the histological and radiological finds of 180 VCF, calculated that the IVC is suggestive of a vertebral osteonecrosis with a sensitivity of 85% and a specificity of 99%. However according to some authors the cleft would not correspond to a necrosis but to a pseudoarthrosis [6,45] or a gas leak from the disc [22]. Given that the cleft sign is most likely an osteonecrosis indicator, it can also be found in other conditions involving a vertebral necrosis, as cancer [44,46,47], osteoporosis [16], radiotherapy sequences [16,44], intraosseous disc prolapse [22,48] and arteriosclerotic vascular disease or alcoholism [34], pancreatitis or cirrhosis [16,37]. Matzaroglou [31] found an incidence of vacuum cleft in patients with osteoporotic VCF selected for vertebroplasty equal to 10-48%. It must be underlined that the lack of IVC does not exclude a KD diagnosis [3].

The bone scan can show alterations in the pathology early stages, before the vertebral collapse takes place, due to the selective build-up of radiolabelled tracer in the damaged vertebra [49]

As there aren't any pathognomonic signs of KD, it is traced back through an exclusion diagnosis, recognising the clinical history as well as the radiological finds, discarding through instrumental and blood chemistry tests, all those causes that could determine necrosis and vertebral collapse.

Histopathology

- The collapsed vertebra histology shows avascular necrosis as well as processes of diffused ossification.
- Avascular necrosis is made up of four concentric areas: a central one made up by devitalized cells, then one that suffered an ischemic stroke, surrounded by an area of hyperaemia and oedema, around which there is healthy tissue.

Few reports are available concerning the histological framework deriving from vertebras suffering a deferred collapse. One of the first ones is supplied by Cardis [50], referred to an L2 body post-mortem evaluation, that macroscopically appeared in the shape of a wedge with haemorrhagic areas in the trabecular bone, while the microscopic exam on the third middle of the vertebral body showed an atrophic degeneration. Afterwards there was the description of some inflammatory alterations, fibrosis of the back, and multiple microscopic fractures, predisposing to the vertebral crush [14]. Several exams targeting the area showing avascular necrosis allowed establishing that this is constituted by four concentric regions [51], a central one made up by devitalized cells, and then one that suffered an ischemic stroke, surrounded by an area of hyperaemia and oedema, around which there is healthy tissue. It was furthermore noticed that inflammatory characteristics such as oedema and inflammatory cells infiltrate can be found only in the disease early stages, while during its advanced stage we can find distributed ossification and fibrosis [5,39]. Finally, in

several cases, the analysed vertebras showed osteoporotic rarefaction, probably because this represents a basic predisposing factor to small fractures that later on evolve in collapse and vertebral necrosis [7].

Treatment

- At present there isn't a standardized therapeutic protocol.
- Non-surgical treatment (bed rest, bracing, lumbar traction, painkillers and anti-osteoporotic medicines) could be considered only in those cases where the vertebral body back wall is intact in neurologically intact patients or in case of severe comorbidity, which contraindicates surgical solutions.
- Surgical therapy aims at treating pain, correcting kyphosis and decompressing the nervous structures
- It is possible to use anterior, posterior or combined surgical airways, according to the operator's experience and preferences.
- In patients without severe kyphosis and without spinal cord impairment, a valid therapeutic option is made up of percutaneous procedures such as vertebroplasty e kyphoplasty.

Currently, also due to the small number of papers on this pathology, there isn't a standardized protocol for KD treatment [31]. The therapeutic choice must be driven by pain intensity, the development of kyphosis and/or neurological deficits, the quality of the bone to be treated and the presence of comorbidity.

Non surgical treatment foresees bed rest, bracing, lumbar traction, painkillers and anti-osteoporotic medicines [38,52]; Fabbriciani et al. [35] administered an 81 year - old woman an osteoanabolic therapy based on *teriparatide*, a recombinant form of parathyroid hormone, obtaining pain resolution, disability reduction and filling of the bone gap. However the use of a conservative management is debated, considering the possible evolution of the pathology in VBC, kyphosis and neurological impairment [53,54] and could be admitted only in those cases where the vertebral body back wall is intact in neurologically intact patients or in case of severe comorbidity which contraindicates surgical solutions [7].

Surgical therapy must be adopted when the pain is unresponsive to conservative treatment or if the patient faces worsening neurological deficits or if there is a progressive deformity of the column in kyphosis and it therefore aims at decompressing the nervous structures and at restoring a correct sagittal balance [55]. The surgical act consists of a vertebral stabilisation or, in case of simultaneous neurological impairment, a stabilisation coupled with a nervous decompression; it must be noted that it is inadvisable to exclusively carry out a decompression without stabilisation as it would result in an additional spine destabilisation and as a consequence an aggravation of the spinal deformity.

Surgical options include a posterior, anterior and a combined approach; furthermore, in selected cases are available minimally invasive percutaneous surgical techniques, namely vertebroplasty e kyphoplasty, notwithstanding that a medullary and/or radicular injury requires an open treatment [56].

The choice of the surgical treatment modality is usually guided by the operator's experience and subjective preferences, but there is the possibility in any case, of referring to classifications leading the decisional process. Mochida et al. [57] divided the finds obtained from radiograms in three types: the first includes a wedge collapse (the height of the vertebral body anterior part is less than 60% of the height of the posterior one) and it would require an anterior decompression with instrumentation, the second type has a concave aspect of the vertebra and the third a flat conformation; it would be preferable to treat these last two types of fracture through a posterior airway. Instead Li [58] takes into account the possibility of executing a percutaneous vertebral augmentation and he identifies three stages, in the first there is a vertebral body compression <20%, in the second a compression >20% and rupture of an adjacent disc, in the third the vertebral body back wall is damaged and a compression of the spinal cord occurs; in the first two stages a simple percutaneous treatment could be feasible, while in the third one it is mandatory the indication to open surgery aimed at nervous decompression and stabilisation.

The anterior procedure enables to obtain a simple decompression with a low risk of affecting nervous structures [59,60], although this entails a longer surgical period and the risk of damaging thoracic or abdominal viscera; excellent results have been obtained by the use of intervertebral tricortical grafts [45] as well as of ceramic glass spacers [60]; in old patients and with poor general conditions, an anterior, transthoracic or retroperiteonal approach would be difficult to tolerate and for this reason a posterior approach is advised.

The posterior procedure allows to effectively decompressing the spinal cord anterior surface, sparing the anterior airway invasiveness, in particular avoiding dissecting the diaphragm [61]. Through the posterior approach is furthermore possible to carry out osteotomies aimed at correcting kyphosis, especially at a low lumbar level.

A possible surgical solution, in patients with a nervous damage, is constituted by the fracture manual reduction with trapedicular insertion of titanium spacer combined with a short segment fixation [62]. According to some authors, the lack of a posterior support would however entail an implant failure, in case of excessive tensile forces [63]; the posterior approach is burdened by a greater risk of damaging nervous structures determining radicular or medullary deficits and fluid fistula [64,65].

An anterior and posterior combined technique would be preferable when a severe sagittal imbalance has been established and it allows to effectively obtaining decompression, kyphosis correction and fusion; a 360° fusion would be particularly recommended in osteoporotic patients [66].

As already mentioned, a valid therapeutic option for amielical patients without severe kyphosis is constituted by percutaneous procedures as vertebroplasty e kyphoplasty, aimed at restoring the vertebral body height but especially at eliminating motility along the clefts, largely responsible for pain symptoms. In order to obtain the maximum stability as regards fractures site, a complete filling with clefts cement is required [67]. The patient must be placed in a prone position in hyperlordosis so to open the cleft and restore the vertebral body height; furthermore cavity-grams should be acquired by injecting contrast medium in the cleft before introducing cement in the vertebra, so to remove the risk of cement leakage [68]. Several studies show how percutaneous procedures are able to relieve the pain and correct deformities [69,70], however they are contraindicated when the back wall is damaged, as retropulsion of bone fragments could occur as well as cement leakage in the spinal canal.

Case Report

In this paper it is presented the case of a patient affected by Kummell's disease which hadn't been promptly diagnosed. Due to the lack of diagnosis the patient's vertebral injury was cured as a simple osteoporotic vertebral collapse; in turn this unsuitable treatment favoured the evolution of the pathology culminating in the compression of the spinal cord and when the patient contacted our hospital he had by now developed a paraparesis.

Case Description

In February 2015, an 81 year-old man was brought to our attention affected by osteoporosis, chronic bronchitis, chronic hepatitis C and

hypertensive heart disease. At the beginning of January 2014 he had suffered an accidental domestic fall on the pelvis experiencing thoracic-lumbar pain lasting for about one month and treated with painkillers; furthermore the patient showed the report of a spinal x-ray performed at the time, which excluded post-traumatic bone injuries. About 4 months after the fall, he suffered a back pain resurgence and another x-ray was carried out showing a reduction in height of the T11 soma, where it was appreciated an opaque strip most likely corresponding to a gap filled with gas (Figure 1); the practitioner that the patient had contacted, diagnosed him an osteoporotic vertebral crush, not considering the cleft signal and he just administered painkillers as well as advising a lumbar belt to be worn in upright posture. In our opinion, in this phase where were still lacking neurological signs and the column deformity wasn't severe yet, it could have been possible to prevent the pathology unfavourable evolution through a vertebroplasty or kyphoplasty. Due to the persistent pain, after 5 months, the patient underwent an MRI, which showed the T11 somatic collapse with oedema, highlighted by STIR sequences, and of low intensity signal wide area on T1 as well as on T2-weighted images (Figure 2). This delayed vertebral collapse together with osteonecrosis highly suggested KD and it should have induced to consider the hypothesis of an open surgical treatment or at least a percutaneous one, however even the second practitioner, probably ignoring the existence of KD, and therefore the relative potential consequences on spinal stability and neurological functions, had classified the fracture as an osteoporotic crush, thus prescribing a pharmacological therapy and again, a lumbar belt. The conservative therapy was hardly effective, but the patient remained amielical until, in January 2015, a progressively worsening strength deficit developed in the lower limbs, until it prevented ambulation. Therefore few days later the vertebral MRI was repeated: the T11 vertebral body appeared even more destroyed and this time, apart from osteonecrosis, there were also gross reparative bone phenomena and a significant compression of the spinal cord (Figure 3). The patient underwent also a bone scan, which showed a selective build-up of radiolabelled tracer at T11 level.

Diagnosis

Only when the patient precipitated in such state, he was addressed to our clinic. He was immediately hospitalized. The patient's personal documentation enabled to reconstruct the typical evolution of Kummell's disease: the initial trauma with corresponding negative x-ray was followed by a period of relative wellbeing, then, after 4 months, lumbar pain reappeared, and this time the x-ray showed T11 collapse accompanied by IVC, suggesting osteonecrosis; the RMI carried out after 9 months from the trauma confirmed the presence of AVN with hypo intensity in T1 and in T2 referable to IVC; after another 2 months, due to the appearance of paraparesis, another RMI was repeated which this time corresponded to the disease final stage, indicating skeletal sclerosis involving stenosis of the spinal canal. Blood chemistry exams were requested (tests for infection, metastasis and multiple myeloma) which excluded other causes of vertebral necrosis. On the basis of anamnestic, laboratory and radiological data, it was reached the diagnosis of Kummell's disease.

Treatment

As there was a medullary compression it was quite urgent to carry out a surgical decompression, simultaneously a vertebral stabilization was indicated. The posterior approach was chosen because the patient, being old and affected by comorbidity, would have hardly tolerated an anterior approach. The spinal cord was decompressed through a laminectomy of T11, while stabilization was obtained introducing some tranpedicular screws in T10 e T12; during the operation were also carried out some biopsy samples through trocars introduced through both the T11 peduncles, in the body of the vertebra.

The histology outcome indicated the presence of necrotic tissue and areas of calcified fibrous tissue.

Outcome

The post-op course was regular and the patient was addressed to a rehab centre, after 5 days of hospitalisation.

One month after the operation, the patient underwent a check-up from which, compared to the pre-operating framework, there was a slight improvement of the strength in the lower limbs and showed x-ray of the spine, which excluded mobilisation of the stabilisation system.

12 months after the operation, after further outpatient and home-based physiotherapy, a follow-up CT was prescribed (Figure 4), which excluded a worsening of the kyphosis level that had been registered before the surgical treatment as well as a further restriction of the vertebral canal. At the same time a clinical exam was carried out, where an improvement of the strength in the lower limbs was found as to enable walking, although with a bilateral support; there still was a modest spinal pain so the patient took painkillers when needed.

Discussion

Kummell's disease consists in a delayed post-traumatic vertebral collapse most likely determined by an avascular osteonecrosis. It isn't known the time range which elapses between the trauma and the insurgence of vertebral necrosis yet, nor the entity that must characterize such trauma. This pathology isn't well known by doctors yet and often in the most important orthopaedics texts isn't even mentioned [71]; for this reason they are only few reports in literature, although it is estimated that, in vertebral fractures, it actually has a high incidence, oscillating between 7% and 37% [4], especially among old patients where the incidence of osteoporosis is very high, which constitutes a fundamental predisposing factor; for this reason, with the world ageing population, the frequency of KD is destined to grow.

A suitable knowledge of Kummell's disease enables to pick the diagnosis and prepare the right treatment. It must be however remembered that there aren't any standardised instrumental or

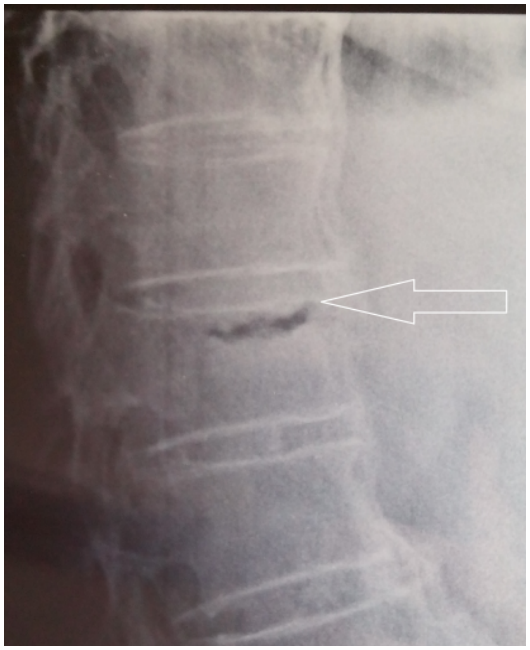

Figure 1: Radiogram after 4 months from the trauma, D11 vertebral body, reduced in height, presents a radiolucent strip corresponding to the sign of the intervertebral vacuum cleft.

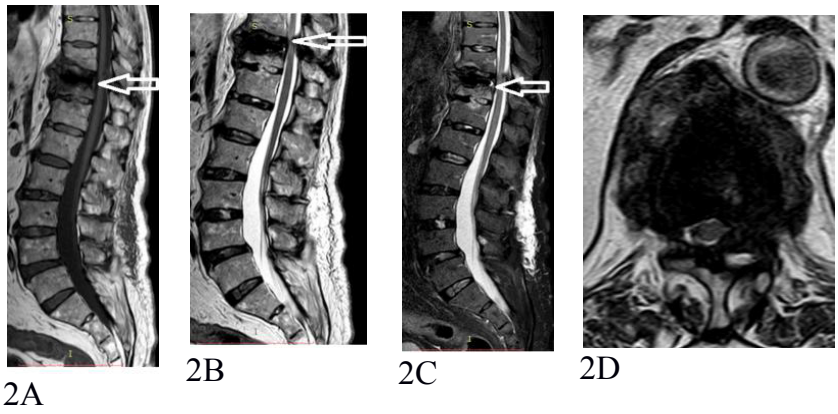

Figure 2: MRI carried out 9 months after the fall, the T11 body has collapsed and it shows a low signal area in the sagittal sequence T1 (2A) and T2 (2B), while the sequences STIR highlights a cancellous bone inflammation (2C); from the axial scan is more evident the fibrous reaction surrounding the necrotic area (2D).

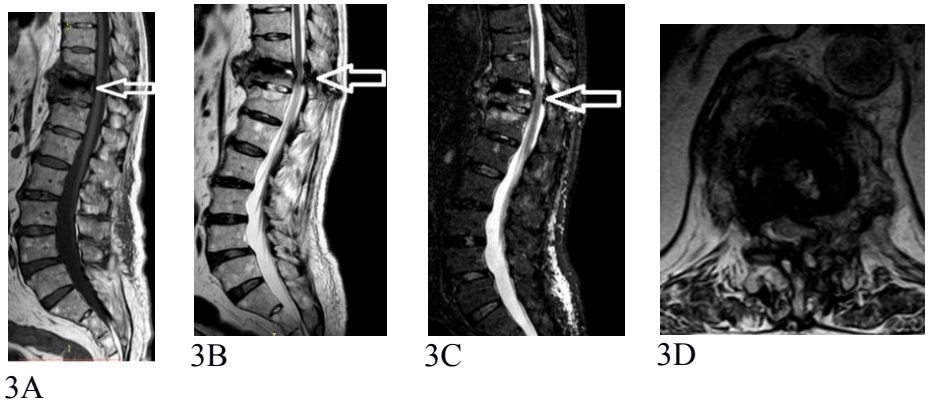

Figure 3: Radiological aspect 1 year after the trauma; MRI with sagittal sections T1 (3A), T2 (3B) and STIR (3C) and in axial section (3D). The development of a deformity in kyphosis, an exuberant bone reaction surrounding the collapsed vertebra and a vertebral canal stenosis with medullary compression at T11 level are obvious.

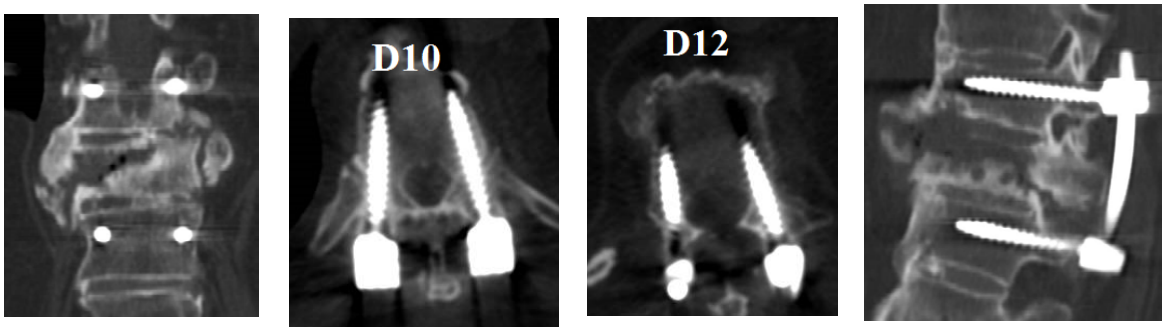

Figure 4: CT 12 months after the operation, with coronal, axial and sagittal reconstructions: a worsening of the kyphosis level registered before the operation as well as a further restriction of the vertebral canal are excluded.

laboratory parameters leading to a diagnosis of KD, therefore a correct diagnostic course foresees the exclusion of other conditions leading to a vertebral necrosis in the absence of a recent and severe traumatic event, in particular of infective and tumour processes.

The typical clinical presentation foresees a spinal pain post-traumatic phase followed by an asymptomatic interval and finally there is the resurgence of pain associated to the column deformity with or without neurological deficits, instead a simple osteoporotic vertebral crush has a more benign course and usually it isn't accompanied by a neurological damage.

An x-ray immediately carried out after the trauma is generally negative but when after few months the symptoms recur radiological exams (X-ray, CT and MRI) show the vertebral body collapse, osteonecrosis, air within the vertebra (cleft signal). The CT highlights in the best possible way the intravertebral vacuum cleft and osteonecrosis, while the MRI finds are characterized by the *double line sign*, namely a low intensity linear region in T1 and in T2, corresponding to the vacuum cleft, which in T2 is surrounded by a high signal area corresponding to an inflammatory reaction [6]; furthermore the magnetic resonance allows a differential diagnosis with pathologies such as spinal cancer and spondylodiscitis [72].

It hasn't even been codified an ideal therapeutic protocol. The

conservative therapy usually isn't effective [9] and it should be reserved to patients who can't undergo a surgical operation due to compromised general conditions. Regarding the surgical treatment it can use minimally invasive techniques, as for example vertebroplasty and kyphoplasty, provided that there aren't any neurological deficits or open procedures with an anterior, posterior or combined anterior-posterior approach, and the choice among such options is basically led by the surgeon's experience.

Conclusion

Kummell's disease should always be suspected in patients, especially if old and with risk factors such as osteoporosis and chronic steroids consumption, who complain about persistent spinal pain after a slight trauma and with a negative radiographic find; these patients should be followed in time with serial radiographic exams. When precocious vertebral necrosis signs such as intravertebral vacuum cleft are found, it isn't advisable to continue a conservative therapy, but on the contrary a percutaneous or open surgical treatment should be carried out, aiming at stopping the course of the disease, which is usually malignant, leading in most cases to severe kyphosis and/or neurological damage. The clinical case brought to our attention shows how an omitted diagnosis implies insufficient therapeutic measures, thus determining devastating repercussions on the patient, both under the clinical and neurological profile; the experience we presented stresses the importance of a satisfying knowledge of KD by the medical world, currently widely ignored, so to implement a therapeutic conduct able to arrest the evolution, often unfavourable, of the disease.

Conflict of Interest

Authors have no conflict of interest to disclose.

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