Thoracic Spinal Tumoral Calcinosis

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ABSTRACT

PURPOSE: Tumoral calcinosis (TC) is a pathological condition that is not well understood. The diagnosis of spinal involvement is possible only with histopathologic examination, and preoperative radiological diagnosis is not possible yet. In this study, a case that was diagnosed with spinal tumoral calcinosis is reported and the related literature reviewed.

CASE REPORT: A 43-year-old male presented with pain in the back and lower back and weakness in the legs. He was paraplegic, had intact deep sensation, and had no incontinence. He was hypo-aesthetic under the level of Th10. DTR and KCR were not taken, and TCR responses were bilaterally indifferent. He had no familial disorders and laboratory tests gave normal results. In the thoracic MRI, a hypo-intense epidural lesion was observed compressing the spinal cord in the T1- and T2-weighted sections at Th8/9 level. The cord also had a signal suggesting edema. The lesion was excised in total with a posterior approach. The Pathology Department evaluated the lesion as calcinosis. The neurological picture improved and recurrence was not noted at the five-year follow-up.

CONCLUSION: TC should be considered in case of lesions with spinal epidural location that are markedly hypo-intense in MRI in T1- and T2-weighted sections and possible surrounding contrast substance retention. TC should be considered in the differential diagnosis of spinal cord compression and constitutes a treatable cause of paraparesis. The treatment is total excision and surgical treatment in the early period is successful.

KEY WORDS: Tumoral calcinosis, Thoracic spine, Paraplegia

INTRODUCTION

Tumoral calcinosis (TC) is a separate clinical and histological entity characterized with periarticular “tumor-like calcium deposits”. In the active phase, there are amorphous and granular calcified material, and mono- and multinuclear macrophage proliferation, osteoclast-like giant cells, fibroblasts and chronic inflammatory elements are found around the lesion. It was first described by Inclan (7). It is most frequently seen in areas surrounding the hip, shoulder and elbow. In the clinic, it is seen dialysis patients in familial fashion or sporadically. The familial type is seen after 20 years of age. It accompanies vitamin D metabolic disorder and mostly affects the region around the hip and the elbow joints (11,18). The treatment is total excision and surgical treatment in the early period is successful. Recurrence rate is high if residues are left back. A diet poor in calcium and phosphate-binding antacids is used for the medical treatment.

Case: A 43-year-old male was urgently operated with a lumbar procedure following the lumbar MRI examination by a neurosurgeon, to whom he had applied with marked and progressive loss of strength in his legs, particularly on the right side and with back and lower-back pain two and a half months ago. His neurological status worsened after the operation. He left the hospital,
which he entered walking, on a wheelchair. When seen by us, he was paraplegic in the motor examination, deep sensation was present in the sensory examination, and he was hypo-esthetic under the umbilical level; the sensory level indicated the T10 level. Deep tendon reflexes and abdominal cutaneous reflexes were unresponsive. The sole cutaneous reflex was unresponsive bilaterally. He had no urinary and anal continence. He had mild back and lower-back pain. All the laboratory test results were within normal limits, including serum calcium, phosphorous, creatinine, alkaline and acid phosphatase and parathyroid hormone levels. One epidural lesion was found in the thoracic MRI in the T8-T9 level compressing the spinal cord. (Figure 1). This pathology was observed as hypo-intense in T1- and T2-weighted sections. The spinal cord also had a signal suggesting edema rather. The patient was operated with these findings. Posterior media approach was applied in the prone position. Following the determination of the level under scopy, approach was made through a 5cm-incision. Laminectomy was performed, and the epidural mass surrounding the spinal cord circumferentially, lobular in structure and detrital-chalky consistence was seen. The lesion was totally excised micro-surgically. A definite gross diagnosis was not possible for the excised pathology. The nodules of the granular calcified deposits divided with fibrous septa consisting of mononuclear inflammatory cells, histiocytes and multinuclear giant cells were seen histopathologically (Figure 2). There was no other proliferative cell, infectious agent or a focus of necrosis in the lesion that the calcification was not seems to be secondary. All of these findings, the lesion was diagnosed as tumoral calcinosis. No complications were seen in the postoperative period. Movement in the legs started after the first week. He

Figure 1: Sagittal and axial MR images: Epidural mass lesions surrounding the spinal cord at T8-9 level, hypo-intense in T1- and T2-weighted sections, compressing the spinal cord and hyper-intense in the T2-weighted sections.

Figure 2: Histopathological examination: A) Calcified nodules divided with fibrous septa, (Hematoxylin -Eosin, x40), B) Lymphocytes, histiocytes and multinuclear giant cells around the calcified deposits (Hematoxylin -Eosin, x200).
was referred to rehabilitation clinic one month later in a condition allowing walking with supports. Neurological deficits were completely resolved after 6 months. The patient that is a farmer is currently working actively, and no recurrences were seen in the 5-year follow-up visit.

**DISCUSSION**

The etiology of this entity is still unknown and no general agreement exists regarding its pathogenesis (5,15,22). In a review by Smack et al (17), 121 cases of TC were identified. They suggest three pathogenetically distinct subtypes: 1- Primary normophosphatemic TC without metabolic abnormalities, with no evidence of familial patterns. Nearly half of the patients live in tropical regions of the world. 2- Primary hyperphosphatemic TC with strong familial patterns. 3- Secondary TC with concurrent disease capable of causing soft tissue calcification as chronic renal failure, hyperparathyroidism, hypervitaminosis D, scleroderma, pseudoxanthoma elasticum, malignancy and milk-alkali syndrome (4) Our patient had no family history, biochemical abnormalities or underlying medical condition known to promote soft tissue calcification. For that reason, we have considered this case as idiopathic or primary TC type1 of the Smack classification.

Spinal involvement was first reported by Riemenschneider (16). Their case was a 59-year-old female, and was diagnosed with a calcified mass involving the L5 vertebral and sacral roots. The largest series of spinal involvement belongs to Durant with 21 cases (3). Spinal TC was found in the x-rays in none of the cases in this series. The lesion in each of the cases was seen in the MRI, however, in the differential diagnosis, pathologies other than TC (tumor, osteomyelitis, discitis, disk hernia, synovial cyst and pannus) were considered. Likewise, meningioma and synovial cyst were mentioned in the differential diagnosis intraoperatively; however, TC was not considered. In this series also, lumbar involvement was the most frequent, and cervical, thoracic, and foramen magnum involvements followed. TC was observed as extradural mass 2-5cm in diameter and related with the vertebral body or posterior elements. The most frequently seen signal pattern is reduced T1 and T2 signals. Our case had a thoracic location with similar features. It was not pre- or perioperatively, and it was only understood with histopathological examination that the mass was tumoral calcinosis.

The gross pathological appearance is characteristic, and is defined as a yellowish mass with lobulated contours in detrital-chalky consistence. There may be cystic areas including yellowish-white fluid in places. In the microscopy, histiocyte and giant cell reaction surrounding the calcium deposits in amorphous plaque fashion. Hydroxyapatite and calcium pyrophosphate hydroxyhydrate crystals were seen together in two cases (3). Contrary to Durant, (14) reported that they observed TC findings in the c-rays in their case. Iglesias (6) found TC lesions at T12 level in a 59-year old patient and at L5-S1 level in a 55-year old patient. The lesion was hypointense in both cases on MRI with minimal circumferential contrast retention, and was diagnosed with synovial cysts radiologically.

TC has been reported for patients in a wide range of ages from infants to adults (1,9,12,19-21). The entity defined as calcific spinal pseudotumor in the literature is thought to be the same disease as TC (10). Total surgical excision of the calcium deposits seems to be an adequate treatment in patients with unknown origin of the disease, as is described in the literature (4,8,9,13,14,18,19). Incomplete excision leads to recurrence. In our case, no regrowth was seen in neuroimaging at the five-year follow-up. The surgical approach can be anterior and/or posterior according to the region and location of the lesion.

A thoracic tumoral idiopathic calcinosis case that was sather similar to our case was reported by Flores et al. (4) with the comment, “In conclusion, TC should be considered in differential diagnosis of spinal cord compression and constitutes a treatable cause of paraparesis.”

**CONCLUSION**

Tumoral calcinosis should be considered in lesions with a spinal epidural location that are markedly hypo-intense on MRI with possible surrounding contrast substance retention. The presence of calcification on CT examination can be helpful in the diagnosis. The treatment of idiopathic tumoral calcinosis is surgical total excision.
REFERENCES


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