

Tumoral Calcinosis in the Upper Cervical Spine Causing Progressive Radiculomyelopathy

—Case Report—

Koichiro MATSUKADO, Toshiyuki AMANO, Osamu ITOU,
Fumiaki YUHI, and Shinji NAGATA

Department of Neurosurgery, Iizuka Hospital, Iizuka, Fukuoka

Abstract

A 54-year-old woman with chronic renal failure presented with tumoral calcinosis manifesting as progressive radiculomyelopathy. Magnetic resonance imaging revealed a spinal epidural mass in the C-2 to C-4 levels. The clinical and radiological findings suggested malignant tumor. Resection of the lesion was performed with total C-2 laminectomy and C-3 and C-4 laminoplasty. The symptoms totally disappeared after surgery. The histological diagnosis was tumoral calcinosis. Tumoral calcinosis is a rare tumoral calcium pyrophosphate dihydrate crystal deposition disease which presents as periarticular soft tissue calcification. Tumoral calcinosis should be considered in patients with a mass lesion involving the upper cervical spine and associated with metabolic abnormalities. Surgical excision is the treatment of choice, because this is completely curative without known recurrence.

Key words: calcium pyrophosphate dihydrate crystal, cervical spine, magnetic resonance imaging, tumoral calcinosis

Introduction

Tumoral calcinosis, first described in 1943,⁴⁾ is a periauricular disease of the hip, elbow, shoulder, calcaneus, knee, and finger. Tumoral calcinosis is characterized by focal and/or multifocal periarticular soft tissue calcification and occasionally occurs in the juxta-articular regions of the extremities.^{8,18)} This benign hereditary disorder occurs mostly in young adults, sometimes with familial clustering. Tumoral calcinosis of the upper cervical spine (C1-2 levels) is extremely rare. This condition should be included among the clinical presentations of calcium pyrophosphate dihydrate (CPPD) crystal deposition disease.⁷⁾ The experience of a case mimicking cervical spine neoplasm suggested that en bloc resection is the appropriate procedure given the uncertainty about the natural history of this lesion.¹⁶⁾

We report a rare case of tumoral calcinosis of the atlantoaxial joint caused by progressive radiculomyelopathy associated with chronic renal failure, which clinically and radiographically resembled

cervical epidural malignant tumor.

Case Report

A 54-year-old woman with a 24-year history of chronic renal failure and receiving hemodialysis three times a week was referred to our department with a cervical epidural mass lesion. She had a 2-week history of progressive radiculomyelopathy, severe upper cervical pain, spastic gait, and weakness of the upper extremities. On admission, laboratory examination of serum calcium, alkaline phosphatase, and uric acid levels found values within normal limits and the C-reactive protein test was negative. Hemodialysis had controlled the blood urea nitrogen and serum creatinine levels to within 50 and 5 mg/dl, respectively. Cervical radiography showed mild spondylotic changes and no definitive calcification. Magnetic resonance imaging revealed a spinal epidural mass from the C-2 to C-4 levels posterolaterally, extending to the atlanto-axial joint. T₁- and T₂-weighted images showed an isointense

Received January 15, 2001; Accepted May 22, 2001

Author's present address: K. Matsukado, M.D., Department of Neurosurgery, Kitakyushu Municipal Medical Center, Kitakyushu, Fukuoka, Japan.

and heterogeneous intensity areas in the spinal cord, respectively (Fig. 1), and an irregularly enhanced mass after gadolinium administration (Fig. 2). The preoperative tentative diagnosis was potentially malignant tumor, such as metastasis or primary sarcoma.

The lesion was surgically resected through a mid-line posterior approach. The posterior surface of the cervical dura mater was covered with a yellowish and brittle substance under the laminae. The structure was contiguous with a mass covered with fibrous tissue between the C-1 and C-2 transverse processes. The yellowish and fragile substance was also recognized inside the fibrous capsule. When this substance was removed, a beak-like atlantoaxial joint, and a fibrous capsule thought to be a joint capsule came directly into view (Fig. 3). As much of the substance as possible inside the capsule was removed, and decompression of the spinal cord was

completed.

Histological examination showed randomly arranged calcium deposits, newly formed blood vessels, histiocytes, and foreign body giant cells inside the fibrous capsule. Both acute and chronic inflammations with macrophage infiltration were observed around the calcium deposits (Fig. 4). There were no tumor cells. The histological diagnosis was tumoral calcinosis.

After surgery, the patient made a good recovery from the spastic gait and weakness of the upper extremities. Eventually she was able to drive to the hemodialysis center by herself.

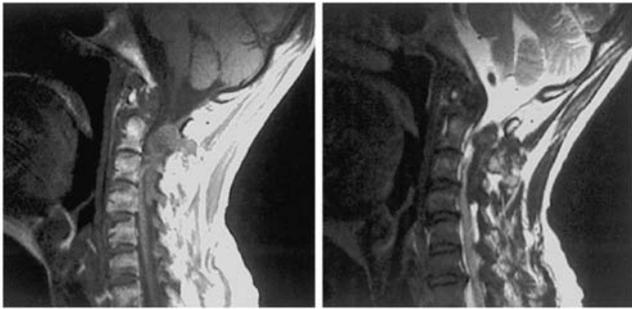


Fig. 1 Preoperative sagittal T₁-weighted magnetic resonance (MR) image (left) showing a mass lesion, isointense to the spinal cord, compressing the spinal cord dorsally, and sagittal T₂-weighted MR image (right) showing the mass as a heterogeneous intensity area.



Fig. 2 Axial post-gadolinium magnetic resonance image showing the irregularly enhanced mass (arrowheads) strongly compressing the spinal cord at C-2 level, and extending anteriorly to the vertebral joint.



Fig. 3 Intraoperative photographs demonstrating the mass encapsulated by thick fibrous tissue between the C-1 and C-2 transverse processes (A), the amorphous and yellowish material inside the tumor (B), and the atlantoaxial joint (asterisks) seen directly after removal of the brittle content (C).

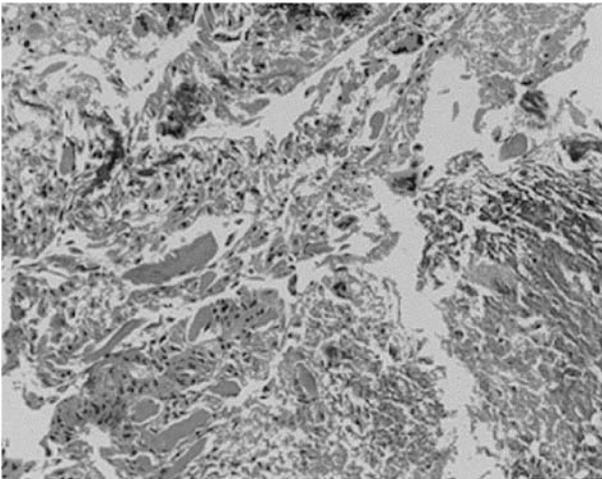


Fig. 4 Photomicrograph of the specimen showing calcium deposits and surrounding newly-formed blood vessels, histiocytes, and foreign body giant cells under the capsule. Hematoxylin and eosin stain, $\times 40$.

Discussion

Tumoral calcinosis of the upper cervical spine is very rare, with only four case reports.^{7,11,14,16} Two cases presented as severe neck pain in adults and two as torticollis in children. No case was associated with compression radiculomyelopathy. CPPD crystal deposition of the spinal ligaments, especially the ligamentum flavum, is well known^{6,12,13,15,19} and induces myelopathy,^{1,2,5} but this ligamentous calcification is considered to be a different entity from tumoral calcinosis. In most cases, the deposits involving the ligamentum flavum were multifocal, relatively small in size, and located in the lower cervical spine. In this case, the mass was completely covered by a fibrous capsule between the C-1 and C-2 transverse processes and proved to be a joint capsule. The capsule content, a yellowish and fragile substance, had spread down to the posterior epidural space and firmly compressed the spinal cord. Ligamentum flavum was identified on the dura in addition to the yellowish and fragile substance. Histological examination showed the thick fibrous tissue forming the capsule appeared to be expanding and was eroded on the interior surface and between its layers by inflammatory reactions.

Laboratory tests of patients with tumoral calcinosis usually reveals no abnormalities or only mild hyperphosphatemia.³ One third of tumoral calcinosis cases are familial with a 50% penetrance, suggesting an autosomal-recessive trait.⁹ Hereditary abnormality of phosphorus metabolism, recurrent

soft-tissue microtrauma, renal failure, and secondary hyperparathyroidism may all be involved in the pathogenesis of tumoral calcinosis. Reduced renal tubular transport of phosphorus and increased renal tubular reabsorption of phosphorus may be factors leading to hyperphosphatemia.¹⁰ The present case was associated with chronic renal failure and long-term hemodialysis.

The natural history of tumoral calcinosis is not well known. The treatment of choice is surgical removal of the lesion when small and amenable to total resection, because recurrence without total removal of the lesion has occurred in other locations.^{17,20} If the total resection is impossible, clinical and radiographic follow up are essential to identify any recurrence.

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Address reprint requests to: K. Matsukado, M.D., Department of Neurosurgery, Kitakyushu Municipal Medical Center, 2-1-1 Bashaku, Kokurakita-ku, Kitakyushu, Fukuoka 802-0077, Japan.