

CERVICAL TUMORAL CALCIUM PYROPHOSPHATE DIHYDRATE DEPOSITION DISEASE 28 YEARS AFTER SUBOCCIPITAL CRANIOTOMY: CASE REPORT

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OBJECTIVE: To describe a rare case of tumoral cervical chondrocalcinosis that appeared 28 years after the patient had undergone suboccipital craniotomy.

CLINICAL PRESENTATION: A 42-year-old woman suffered from cervicalgia associated with a firm mass at the occipitocervical region. Plain x-ray and computed tomographic and magnetic resonance images revealed a calcified lesion in a scar from a previous suboccipital craniotomy.

INTERVENTION: The patient underwent tumorectomy and histopathology, which revealed an exuberant tumoral chondrocalcinosis. Laboratory test results revealed no secondary cause for the chondrocalcinosis.

CONCLUSION: Identification of chondrocalcinosis beyond the cervical region is very rare. Localization of chondrocalcinosis in a scar from a previous suboccipital craniotomy has not been previously reported. Surgery appears to be the treatment of choice for this form of chondrocalcinosis.

KEY WORDS: Calcium pyrophosphate dihydrate disease, Cervical spine, Craniotomy, Tumoral calcinosis

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Arthropathies that result from the deposit of pyrophosphate crystals in soft tissues have been the subject of numerous publications since the early 1960s. The term "chondrocalcinosis" was introduced in 1963 by Zitnan and Sitaj (35); however, their contemporaries, McCarty et al. (20), preferred to call this pathology a "pseudo-gout syndrome." In 1977, after he reviewed the literature on the subject, McCarty (19) proposed the designation "calcium pyrophosphate dihydrate crystal disease." This arthritic disorder is generally found in the elderly, and in the great majority of cases it affects the joints of the pelvis and the limb extremities (24). Neurological deficit or bone lysis occurs when the spine is affected (21, 22).

There are two interesting points concerning this case. First, in this patient we have the only reported case of cervical chondrocalcinosis that includes neither neurological symptoms nor osseous disruption and is associated with intense cervicalgia that was partially relieved by surgical treatment. Second, it is interesting for neurosurgeons to note that the patient's lesion appeared on a scar from a suboccipital craniotomy that was performed 28 years earlier.

CASE REPORT

In 2004, a 42 year-old woman with cervicalgia was admitted to the hospital. Her pain had been radiating to the occipital and scapular areas for 4 months. In this time, the pain progressively worsened and proved refractory to standard analgesics.

In 1976, when the patient was 14 years old, she underwent surgery to relieve symptoms of intracranial hypertension syndrome. This involved exposure of the posterior cranial fossa, which revealed an arachnoid cyst.

A neurological assessment in 2004 yielded normal findings, although it revealed a sensitive and firm swelling located at the occipitocervical junction. Standard x-rays of her spine (*Fig. 1*) showed a flaky, apparently calcified lesion that extended from theinion to the superior border of the spinous process of C4 at the midline. The lesion appeared to develop within the soft tissue rather than the bone structures. Computed tomographic imaging of the cervical spine (*Fig. 2*) confirmed that the structure was calcified in nature. The lesion had a heterogeneous aspect and extended from



FIGURE 1. This sagittal standard x-ray of the cervical spine shows a pseudotumoral calcified lesion behind the spinous process of C1–C2 and C3.



FIGURE 2. Axial computed tomography reveals a calcified lesion within the soft tissue with no indication of bone erosion.



FIGURE 3. A sagittal T1-weighted magnetic resonance image indicates the presence of a lesion within the soft tissue that has no contact with bone (with respect to the epidural space at C1 level).

the suboccipital region to the spinous process of C3 without canal invasions.

The lesion was detected on magnetic resonance images as an isointensity area in T1-weighted sequences (Fig. 3).

In April 2004, the tumor was removed via a posterior approach. The resection of the calcium pyrophosphate dihydrate disease was complete. Surgery revealed a crumbly, whitish tissue that was easily distinguishable but hard to separate from the surrounding muscles. The growth had not penetrated into the region in front of the laminae. Total excision

was accomplished. The anatomopathological assessment revealed large, purplish, crystalline deposits that were identified as calcium pyrophosphate dihydrate crystals in high-magnification microphotographs (square-tipped crystals that were weakly refringent under polarized light). A strong, dissolving macrophage reaction surrounding the deposits was apparent, as was giant multinucleate cell proliferation. We found no sign of malignancy and diagnosed this as cervical chondrocalcinosis.

Standard x-ray images of the wrists, hips, and pelvis showed no other stigma of chondrocalcinosis. Laboratory test results revealed no secondary causes for chondrocalcinosis.

The follow-up duration after the second surgery was 18 months. At our last clinical examination, the patient reported she was free of pain and demonstrated good cervical mobility. Her scar was clean.

Standard x-ray control images did not reveal any calcification.

DISCUSSION

Chondrocalcinosis belongs to the arthritis family because of the inherent role of microcrystalline deposits in the disease (6, 16, 17, 23, 29): calcium pyrophosphate dihydrate crystals deposit within the joints. It is a pathology mostly confined to the elderly population except for certain unusual cases such as hypomagnesemia or Coffin-Lowry syndrome (24). Its prevalence increases with age, reaching close to 45% in subjects over 85 years old (24), generally with no symptomatic manifestation. The most common sites for the chondrocalcinosis include the hip, elbow, shoulder, and foot. In the major-

ity of cases, its onset is diffused and involves more than two joints, usually symmetrically (24).

This pathology of the elderly, which is particularly predominant in females, should be approached with a rigorous initial etiological workup to search for the presence of morbid conditions that are sometimes associated with this disorder, such as psoriasis, hyperparathyroidism, hemochromatosis, hypophosphatasemia, hypomagnesemia, hyperuricemia, Wilson's disease, traumatism, and past history of surgery (4, 6, 8, 24). However, primitive and idiopathic forms of chondrocalcinosis are the most frequently encountered; family-related forms are uncommon.

Chondrocalcinosis is a risk factor for neck pain. It frequently involves the cervical spine, and as such, may be associated with development of neck pain. This diagnosis should be considered when a patient presents with neck pain (9).

For this patient, surgery was the logical indication; she had a mass within a previous scar, and her clinical examination revealed abnormal findings. The clinical and radiological data showed strong correlation. Surgery was important, both to confirm the anatomopathological assessment and to relieve her clinical symptoms.

Localization of calcium pyrophosphate dihydrate crystals in joints is most common, but extra-articular deposits have also been reported to occur, within the ligamentum flavum (1, 3, 7, 18, 19, 20), the dura mater (12, 14), and the tendon calcaneus (10). The presence of such intra-articular deposits does not necessarily cause symptoms that are either clinically or radiographically recognizable (24), and clinical symptoms can occur that do not show in x-rays. These conditions often render the diagnosis somewhat tricky.

Spinal localizations of these crystals have rarely been reported in the literature. Those that have been reported describe cervical (2, 11–13, 15) and dorsal medullary compression (26), lumbar sciatica (22, 27), and pseudospondylolisthesis (30). They are extra-articular and lie within the ligamentum flavum, posterior common ligament (23, 28), atlanto-occipital ligament (5), vertebral body (21), and annulus fibrosus of intervertebral discs (21). Such deposits are always nodular and of low volume, but because they are located within the spinal canal, they can induce cord compression and neurological deficits. All vertebral levels are involved, from the foramen magnum (5, 27) to the lumbosacral hinge (22). Cervical localizations occur with the highest incidence. In 1993, Baba et al. (1) reviewed 91 cases of cervical myelopathies, to which he added 8 personal cases. According to him, 8 times out of 10, this myelopathy has a slow and gradual evolution, but it can suddenly decompensate after minor trauma. Baba et al. confirmed the predominance of this condition in females, as it affected 84 women and only 15 men. A tumoral form similar to that in our patient was described only once, by Kokubun et al. (16): they reported the case of a 68-year-old woman who suffered from cervicalgia associated with restrained motion of the cervical spine in all directions. Radiographic evaluation and intervention revealed a calcified, multilobular, pseudotumoral

lesion located in the interlaminar portions of C1 and C2. During surgery, the lesion proved to be very adherent to the dura mater. More chondrocalcinosis nodular deposits were found in the ligamentum flavum at the same level, which indicates that the ligamentum flavum may have been the point of origin.

CONCLUSION

Our observations are interesting because they report an unusual localization of cervical chondrocalcinosis: it touched neither the dura mater nor the ligamentum flavum. That this tumor was strictly extracanalicular accounts for the unusual clinical manifestations that we encountered, including the existence of considerable pain in the patient with no sign of spinal cord compression, and the patient's positive response to surgical treatment.

The link between a preexisting scar and this kind of tumor has already been described, but it is not completely understood.

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COMMENTS

Scavarda et al. report a rare case of calcium pyrophosphate deposition disease (pseudogout) that caused cervicalgia in a patient who had suboccipital craniotomy 28 years earlier. The lesion is rare because it arose within a scar of a previous surgery rather than within a more typical spinal location, such as the ligamentum flavum. Spinal surgeons should note that this lesion was readily visible on a lateral cervical x-ray, and its removal can potentially relieve neck pain in affected patients.

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Scavarda et al. describe a most unusual case of a woman who developed a symptomatic mass in the posterior cervical region, which was ultimately found to be consistent with calcium pyrophosphate. The patient responded well to excision of the mass. This case is also unique in that this patient had a posterior fossa craniectomy 28 years before the development of this mass. Whether the previous surgery was related to the calcium pyrophosphate deposition is unclear.

Surgeons should be aware that soft-tissue deposition of calcium pyrophosphate may occur, and those patients who are symptomatic may well respond to surgical resection, as did this patient.

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Scavarda et al. present a rare case of calcium pyrophosphate deposition. Its association with previous surgery is most certainly suspect from an etiological perspective. This patient fits into the archival category of rare cases. It is important to keep this imagery in mind when dealing with similar patients in the future.

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