

Thoracic Myelopathy from Coincident Fluorosis and Epidural Lipomatosis

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Skeletal fluorosis is rare in North America. It can present with back pain and extremity weakness. Immobilization of the spine and the extremity joints can occur. It is usually caused by abnormally increased oral fluoride intake over many years. Epidural lipomatosis is usually caused by idiopathic obesity or corticosteroid use. It has been linked to highly active antiretroviral therapy (HAART) for human immunodeficiency virus (HIV). Fluorosis and epidural lipomatosis are each rare causes of compressive myelopathy, and have never been described previously as a combined cause of spinal stenosis leading to myelopathy. We describe an unusual case of thoracic myelopathy due to coexistence of both conditions.

CASE REPORT

A 46-year-old man presented with subacute onset bilateral leg weakness and parasthesias over one-year associated with low back pain. He progressed to wheel chair dependence, only able to ambulate short distances with a walker. His past medical history was significant for HIV. He had been treated for over eight-years with HAART, and his current regimen consisted of emtricitabine, tenofovir, and efavirenz. His CD4 count was $420 \times 10^6/L$ with an undetectable viral load. The patient also had anxiety treated with paroxetine and lorazepam. Psychiatric evaluation revealed significant chronic benzodiazepine intoxication and dependence with daily doses of lorazepam ranging from 80-140 mg daily. In addition, he was diagnosed with pica and admitted consuming up to six tubes of toothpaste weekly since childhood. He was not from and had never visited a region endemic for fluoride toxicity. Cognitive impairment due to benzodiazepines precluded valid assessment of obsessive, mood, or psychotic causes for his toothpaste ingestion.

Neurological examination revealed spasticity in the lower extremities with 4/5 power in a pyramidal distribution and hyperreflexia. There was a sensory level at T8-T9. Vibration sense was impaired to the mid shins, joint position sense was absent at the toes, and the Romberg sign was positive. The patient had impaired heel-shin testing in keeping with his motor and sensory impairment, and his gait was wide-based. General examination showed stained and pitted teeth. He was mildly obese.

Serum fluoride was $44.2 \mu\text{mol/L}$ (normal range 1 to $4.6 \mu\text{mol/L}$).

Bone mineral density (BMD) was markedly increased. The mean lumbar spine (L1 to L4) BMD was 2.498 g/cm^2 for a T-score of 10.6 and a Z-score of 10.8. The total proximal femoral BMD was 2.670 g/cm^2 for a T-score of 12.2 and a Z-score of

12.5. These values indicate that the BMD in the lumbar spine and proximal femur were over 10 and over 12 standard deviations above the mean of normal young-adult controls and age-matched controls, respectively.

Plain radiographs confirmed profound sclerosis of the spine and femurs, as well as ribs, pelvis, and sacrum.

Bone marrow biopsy demonstrated severe cortical and trabecular sclerosis with preserved lamellar structure and prominent osteoblastic activity consistent with skeletal fluorosis. Hematopoietic tissue was markedly reduced but showed orderly maturation. No lymphoma, granuloma, or metastases were seen. Anterior longitudinal ligament (ALL) and ligamentum flavum (LF) hyperostosis, as well as diffuse-idiopathic-skeletal-hyperostosis like osteophytes, were seen on computed tomography (CT) (Figure 1). Spine magnetic resonance imaging (MRI) demonstrated diffuse sclerosis of bone which was hypointense on T1- and T2-weighted images (Figure 1). Normal fatty marrow was completely absent. Multilevel anterior bridging osteophytes were seen with an appearance similar to diffuse-idiopathic-skeletal-hyperostosis. There was ALL hyperostosis plus mild thickening and sclerosis of the posterior longitudinal ligament. Hyperostosis of the LF was evident at multiple levels, but was particularly mass-like in the thoracic spine. There was also marked thoracic epidural lipomatosis, most prominent at levels T5-T9. The combination of LF hyperostosis and epidural lipomatosis filled much of the spinal canal at T5-T9. There was resultant severe narrowing of the thecal sac with cord flattening and subtle T2 hyperintensity in the cord at T7-T8.

The patient opted for neurosurgical management which included T5-T9 laminectomy. Considering the brittle nature of hyperostotic bone, a wide laminectomy was performed. The LF was noted to be significantly ossified and necessitated the use of a high speed drill for its removal. The underlying epidural fat was resected using cautery and dissection. Intraoperative

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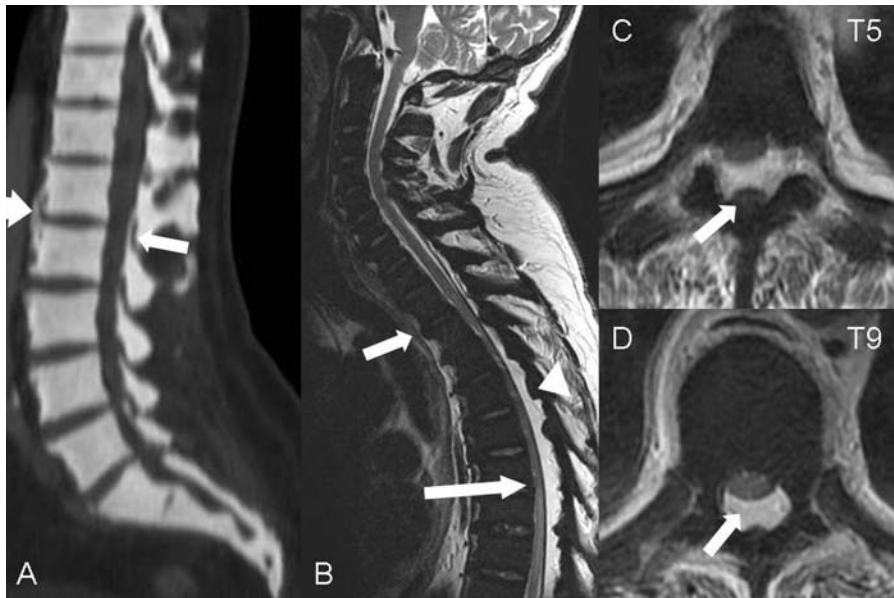


Figure 1: Pre-operative sagittal CT lumbar spine (A); sagittal T2 MRI cervical thoracic spine (B); and axial T1 MRI thoracic spine (C, D). Sagittal CT lumbar spine (A) demonstrates severe osteosclerosis, anterior longitudinal ligament hyperostosis (A, short arrow), and ligamentum flavum hyperostosis (A, long arrow). Sagittal T2 MRI of the cervical thoracic spine (B) demonstrates diffusion marrow hypointensity compatible with osteosclerosis, anterior longitudinal ligament hyperostosis with large anterior bridging osteophytes (B, short arrow), and ligamentum flavum hyperostosis (B, arrowhead). There is thoracic epidural lipomatosis, most severe at T5 to T9, with resultant severe thecal sac narrowing, cord flattening, and subtle T2 hyperintensity in the cord at T7-T8 (B, long arrow). Axial T1 images of the thoracic spine (C, D) demonstrate severe ligamentum flavum hyperostosis (C, arrow) and severe epidural lipomatosis (D, arrow) with resultant severe thecal sac narrowing.

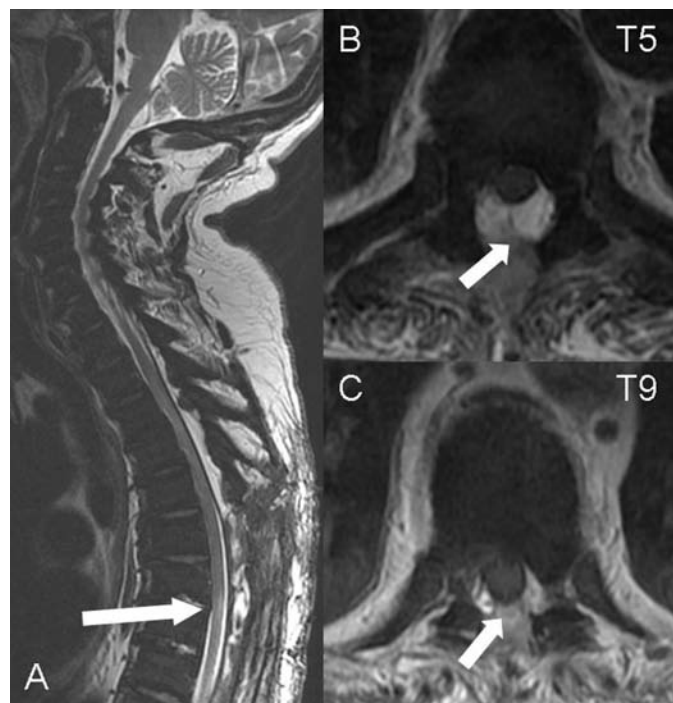


Figure 2: Post-operative sagittal T2 MRI cervical thoracic spine (A); and axial T1 MRI thoracic spine (B, C). Sagittal T2 MRI of the cervical thoracic spine (A) demonstrates an extensive posterior bony decompression with resection of the thickened ligamentum flavum and epidural lipomatosis. The thecal sac is no longer compressed but there remains a small focus of T2 hyperintensity in the cord at T7-T8 (A, arrow). Axial T1 images of the thoracic spine demonstrate that the thecal sac and cord are decompressed, and now have with more circular shape (B and C, arrows).

findings suggested that the non-flexible nature of the ossified LF resulted in compression of the increased epidural fat and underlying spinal cord. The patient's post-operative course was uncomplicated, and he was discharged to a rehabilitation facility. At three-months after surgery, examination revealed normal tone and power except for 4+/5 power of the right knee flexors and dorsiflexors. There was no hyperreflexia or sensory level. He was ambulating approximately 200-meters with a cane. He denied further toothpaste consumption. Follow-up MRI demonstrated good decompression of the thecal sac and cord (Figure 2).

DISCUSSION

Fluorosis can occur from over exposure in food, water, air, and excess use of toothpaste¹⁻³. Most cases involve chronic exposure to elevated fluoride levels, exceeding 10 mg daily. Our patient consumed up to 100 mg of fluoride daily since childhood. Skeletal fluorosis is rare in North America where most of the population lives in urban centers and have access to drinking water with well controlled fluoride levels. However, increased fluoride levels can be seen in North Americans with access to well water only⁴. The disease is endemic to India, China, and parts of Africa where fluoride levels in drinking water are poorly controlled. Elevated fluoride levels can be detected in the blood, urine, and nails of those effected¹.

The most striking feature of skeletal fluorosis is diffuse osteosclerosis. There is frequent forearm interosseous membrane hyperostosis. Osteoarthritis, osteonecrosis, and fractures can occur as a result of the marked bony hardness. Some bone changes can imitate a seronegative arthritis⁵. Ligamentous hyperostosis occurs. Hyperostosis of the spinal ligaments, particularly the LF, can lead to compressive myelopathy or myeloradiculopathy⁶⁻⁹.

Symptomatic epidural lipomatosis is usually idiopathic but has been associated with severe obesity, endogenous or exogenous corticosteroids, and hypothyroidism¹⁰. A complication of HAART is HIV lipodystrophy, consisting of abnormal fat distribution with metabolic disturbances including insulin resistance, deranged glucose and lipid metabolism¹¹. Human immunodeficiency virus lipodystrophy can be seen with all classes of HIV antiretroviral therapies. Epidural lipomatosis can result¹²⁻¹⁶.

This patient had been on HAART for over eight years. He did have severe epidural lipomatosis, probably related to both HAART and mild obesity. Hyperostosis of all spinal ligaments, particularly the ALL and LF, was seen. The LF hyperostosis in combination with epidural lipomatosis resulted in severe thoracic thecal sac narrowing. T2 signal change was seen in the cord at T7-T8 consistent with myelomalacia. This explained the patient's lower leg weakness.

The treatment of myelomalacia associated with LF hyperostosis is posterior spinal decompression. This treatment has had mixed results^{7,8}. Decompression has also been used in cases of symptomatic epidural lipomatosis¹⁷⁻¹⁹. Our patient decided to proceed with surgical decompression and had a good clinical outcome.

CONCLUSION

We present a case of skeletal fluorosis due to chronic toothpaste pica and epidural lipomatosis probably related to

HAART and mild obesity. There was resultant severe thoracic thecal sac narrowing and symptomatic myelomalacia. Both conditions have rarely been independently reported to cause compressive myelopathy. The presence of both in one individual with myelopathy has not been previously reported to our knowledge.

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