Patients with scleroderma, otherwise known as progressive systemic sclerosis, usually present with one or more components of the “CREST” syndrome, which includes calcinosis, Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia. Calcinosis refers to soft-tissue calcifications that are seen as a late manifestation of progressive systemic sclerosis in 9 to 27% of cases. Rarely, calcinosis is manifested as a large tumor-like mass that may cause symptoms by impinging on contiguous structures. The calcinotic lesions associated with progressive systemic sclerosis are usually seen in the finger tips and around the synovial joints of the hands, knees, and elbows. Rarely, calcinosis is manifested as a large tumor-like mass that may cause symptoms by impinging on contiguous structures. Paraspinal calcinosis has been the subject of a few case reports; however, its occurrence in the cervical spine with attendant canal compromise and spinal instability has not been discussed. We report a case of symptomatic cervical paraspinal calcinosis in a patient with progressive systemic sclerosis and discuss the cause and treatment of this disorder.

Case Report

This 65-year-old woman was diagnosed with progressive systemic sclerosis 10 years prior to neurosurgical consultation. The manifestations of her disease consisted of sclerodactyly and esophageal dysmotility, and the symptoms and signs were mild and nonprogressive. Approximately 5 years after the onset of other symptoms, the patient began to complain of neck pain. This pain was mostly localized to the upper posterior aspect of her cervical spine, with some radiation to the interscapular region. This pain had become significantly more severe as time went by. It was exacerbated by changes in posture, ambulation, and especially by riding in a car. The pain was ameliorated by recumbency and wearing a hard cervical collar. Coughing was noted to cause Lhermitte’s sign. She denied any weakness, numbness, gait abnormality, or problems with bladder or bowel control.

Examination. Physical examination revealed an otherwise healthy, active woman with stigmata of scleroderma. A large, firm, nonmobile, slightly tender mass was palpated in the posterior superior cervical spine region. Neck rotation was limited to 20° in each direction, flexion was limited to 90°, and extension to 60°. Her mental status was normal. Cranial nerve examination was only significant for slight leftward tongue deviation. Her strength and sensation were normal. She was normoreflexive with no Hoffman or Babinski sign.

Lateral cervical spine roentgenograms revealed a large posterior, calcified, lobulated mass extending from the occiput to the top of C-5, with no gross intervertebral movement on flexion–extension (Fig. 1). Computerized tomography (CT) scanning demonstrated that the mass was homogeneous, lobulated, of bone density, and seemed to be centered on the facets (Fig. 2). The mass extended posteriorly into the paraspinal musculature and anteriorly around the left lateral aspect of the vertebral body and surrounded the vertebral artery. Magnetic resonance (MR) imaging and MR angiography were performed to evaluate neural and vascular compromise more fully (not shown). Magnetic resonance imaging revealed the mass to be en-
croaching into the spinal canal but not compressing the cord. Magnetic resonance angiography demonstrated a significant reduction in flow in the left vertebral artery.

The patient had been treated conservatively for 18 months. She was receiving a course of diltiazem, a calcium channel blocker, with the hope of reducing the calcinotic mass. The worsening of the mechanical neck pain (relieved by application of an external orthosis) and the recently developed Lhermitte’s sign on coughing prompted surgical intervention.

Operation. After administration of general anesthesia, the patient was placed prone and positioned in a three-pin head holder. A dorsal midline incision was made, which exposed a large mass (7 × 10 cm) over the posterior aspect of the upper cervical spine. The lobulated mass appeared mostly soft and chalky but there were several cysts filled with milky fluid. The posterior elements of C1–4 were involved by the mass, and the thecal sac was compressed at C2–4. After subtotal removal of the mass, the canal was observed to be satisfactorily decompressed. The spine was then stabilized by placing a contoured titanium rod from the occiput to C-7 and affixing sublaminar cables bilaterally at C5–7. The laminae and facets were found to be soft; thus, the instrumentation was imbedded in methyl methacrylate to provide a solid, immediate fusion. To provide long-term stability an autologous bone graft fusion was performed.

Postoperative Course. The patient’s recovery was uneventful. She was free of mechanical neck pain postoperatively and her Lhermitte’s sign on coughing had abated. Postoperative roentgenograms demonstrated good cervical alignment and placement of instrumentation (Fig. 3). Calcium channel blocker treatment was continued.

At 1-year follow up the patient was free of neck pain and without change in her neurological status or recurrence of Lhermitte’s sign. Imaging revealed good bone fusion and no change in the size of the mass. Microscopic examination of the specimen revealed dense connective tissue with amorphous calcified mineral deposits (Fig. 4).

Discussion

Pathogenesis of Calcinosis

The biochemical mechanisms underlying the pathological calcium deposition in scleroderma are unclear. Several authors have noted a significant association of anticentromere antibodies in those patients who develop calcinosis. The production of anticentromere antibodies occurs in patients with progressive systemic sclerosis who develop an autoimmune reaction to the centromeres in mitotically active cells.

Electron micrographic studies have demonstrated soft-tissue calcinosis in progressive systemic sclerosis to be the result of calcium crystal deposition. Deposits are found mostly in the extracellular matrix of muscle, periarticular tissues, and cartilage. Intracellular deposits are sometimes seen in macrophages and histiocytes.
Paraspinal calcinosis

Fig. 4. Photomicrograph of a specimen of the calcinotic lesion revealing dense connective tissue with amorphous calcified mineral deposits. H & E, original magnification × 100.

Origin of Symptoms

The tongue deviation observed in this case was most likely caused by the entrapment of the hypoglossal nerve some distance from its egress from the skull. Imaging studies showed no abnormality of the hypoglossal canal. The anterior extent of the calcinotic mass, as shown in Figs. 1 and 2, would allow for the entrapment of the nerve in the neck.

In the case reported here, the disease process had compromised the integrity of cervical facet joints over several vertebral levels. This damage was not severe enough to cause overt instability; thus, gross movement could not be demonstrated on flexion–extension films. However, over many months micromotion at the affected area led to irritation of the neural elements, resulting in Lhermitte’s sign and mechanical neck pain. The insidious, progressive syndrome of pain and signs of neural compression resulting from subclinical instability have come to be known as glacial spinal instability.1

Surgical Treatment

The goal of surgery was to decompress the spinal canal to alleviate the patient’s neck pain and Lhermitte’s sign, which we believed were symptoms of instability. Rigid immobilization by means of internal fixation relieved the patient’s symptoms and signs, providing indirect evidence of instability as their cause.

Long-term stability can only be provided by solid bone fusion, which was achieved by autologous bone graft in this case. Methyl methacrylate was used to augment the hardware in providing immediate stability and preventing the wires from pulling through the patient’s unusually soft bone, which would have caused the instrumentation to loosen.

Recent reports suggest that the soft-tissue calcinosis associated with collagen vascular disorders may be treated medically. One group noted the regression of a calcinotic mass over a 2-year period in a woman being treated for hypertension with diltiazem, a calcium channel blocker.1 The authors suggested the therapeutic effect was due to the inhibition of calcium influx into cells. Warfarin, in low doses, has also been used to treat calcinosis.1 The mechanism of action of warfarin in this setting is unclear. The administration of a calcium channel blocker in this case may have contributed to the arrest in growth of the calcinotic mass.

Complete resection of the mass was not attempted for several reasons. An adequate canal decompression and exposure for stabilization were achieved by subtotal resection. A complete resection would have put the encased vertebral artery at risk. We believed this risk could not be justified because the patient did not have symptoms referable to the vertebrobasilar system. The underlying process of tissue calcinosis was addressed medically to prevent further growth of the calcinotic mass and thus avert any future neurovascular compromise.

Conclusions

We present a case of paraspinal calcinosis in a patient with progressive systemic sclerosis. In this unique case, the calcinotic mass was centered over the cervical facet joints at several levels, causing spinal instability and mechanical neck pain. The treatment in this case was chosen to achieve spinal decompression and stabilization with adjuvant medical therapy prescribed for controlling progression of the calcinosis.

References


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