Surgical decompression in spinal Paget's disease: illustrative case

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BACKGROUND  Paget's disease of bone (PDB) is a common bone metabolic pathology in older adults, characterized by mixed osteolytic, osteoblastic, and quiescent periods. Surgical guidelines for PDB involving the spine are not well-defined and are reserved for cases refractory to medical treatments, typically bisphosphonates like zoledronic acid. This case study describes a 52-year-old male with PDB who presented with rapidly progressing myelopathy symptoms refractory to standard medical treatment, warranting surgical decompression.

OBSERVATIONS  Surgical decompression and fusion, involving laminectomy with partial facetectomies, placement of pedicle screw instrumentation, and posterolateral arthrodesis spanning beyond the pathological segment, was performed. Follow-up visits indicated progressive improvement in symptoms and mobility, and imaging showed stable postsurgical changes with increased sclerosis in the affected vertebrae on a 2-year postsurgical course.

LESSONS  This case underscores that PDB of the spine can transition from asymptomatic to significant impairment and demonstrates that surgical intervention can provide effective symptomatic relief in myelopathy secondary to PDB. The case contributes to the growing evidence of the effectiveness of surgical decompression in PDB involving the spine.

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KEYWORDS  Paget's disease of bone; spinal decompression; spinal sclerosis; Paget's disease of bone management; myelopathy

Paget's disease of bone (PDB), named by its discoverer James Paget in 1877, is the second most common bone metabolic pathology seen in older adults. It is characterized by focal regions of accelerated bone remodeling. The progression of the disease through several phases has been described, including an initial increase in osteoclasts (osteolytic), followed by increased osteoblasts (mixed osteolytic-sclerotic), and "burned-out" or quiescent periods with decreased bone cell activity, resulting in a sclerotic radiographic appearance and residual abnormal bone structure.1,2 PDB most frequently affects the bones of the pelvis, spine, skull, femur, and tibia. Although many cases of PDB are asymptomatic, patients can develop musculoskeletal, neurological, cardiovascular, or metabolic dysfunction and have an increased risk for primary malignancies. Specific to neurological symptoms, hearing loss and cranial nerve dysfunction, myelopathy, and radiculopathy due to bony overgrowth around neural structures can occur. Medical therapies including bisphosphonates have been highly effective in normalization of biochemical markers, improvement in pain, stabilization of the disease, and even reversal of neurological symptoms/deficits. Currently, surgical guidelines for PDB are unclear, but previous studies have shown the possible benefits of decompressive spine surgery for medically refractory back pain and neurological dysfunction related to PDB. We present an interesting case of surgical decompression of the spine in a 52-year-old patient with Paget's disease due to rapidly progressing myelopathy symptoms.

Illustrative Case

A 52-year-old male with a history of hypertension and chronic back pain presented with progressively worsening gait dysfunction and altered lower extremity sensation over 1 year. Historically, he had elevated total serum alkaline phosphatase (ALP) levels, and a bone scan 15 years earlier that was suspicious for Paget's disease of the thoracic spine, but no treatment was pursued at that time. His presenting physical examination revealed back tenderness with diffuse, bilaterally

ABBREVIATIONS  ALP = alkaline phosphatase; CT = computed tomography; MRI = magnetic resonance imaging; PDB = Paget's disease of bone.

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decreased sensation from the midthorax and below, mildly increased reflexes in the lower extremities, and a positive Romberg’s sign.

Computed tomography (CT) and magnetic resonance imaging (MRI) scans showed blastic, sclerotic lesions at T7–10, severe spinal stenosis, and raised concerns for PDB (Figs. 1 and 2). A full-body bone scan indicated diffusely increased activity isolated to the mid-lower thoracic spine consistent with localized Paget’s disease (Fig. 3). Laboratory results showed elevations in C-terminal telopeptide of 211 pg/mL, bone-specific ALP 256 µg/L, and ALP 757 U/L. A biopsy of the sclerotic lesion confirmed PDB. Treatment was initiated with zolendronic acid infusion and daily cholecalciferol 2000 IU. Despite the initiation of medical treatment, the patient’s condition rapidly progressed to severe sensory, motor, and gait dysfunction, leading to wheelchair dependency, and new-onset urinary frequency.

Due to the rapidly progressing symptom burden from myelopathy, surgical decompression was recommended. Given the multiple levels of decompression required, the risk for iatrogenic instability due to bony resection, and the weakened structural integrity of the pagetic bone, the patient was also recommended for posterolateral fusion. The extent of fusion was planned to span beyond the pathological bone and ensure multiple solid points of fixation in normal bone not at risk for progression of Paget’s disease or existing structural compromise.

The patient underwent laminectomy of T6–11 with partial facetectomies at T7–10 and posterolateral instrumented fusion of T5–12. At surgery, there was severe spinal stenosis from a combination of hyperostosis and epidural tissue, including a compact layer of fat, ligamentum, and numerous large epidural veins. The sclerotic bone had numerous small vascular channels that were in communication with the epidural veins, and liberal use of bone wax at laminectomy edges, hemostatic agents, and bipolar cautery allowed for hemostasis, but total blood loss for the operation was greater than 3000 mL. Intraoperative ultrasound was performed after completion of the posterior approach with laminectomy and medial facetectomy. Intraoperative ultrasound demonstrated decompression of the spinal cord without residual compression ventrally by the vertebral body or laterally by the bony pedicles; thus, resection of the pedicles or vertebral body was not performed. The patient had immediate postoperative improvement in neurological function, including increased proximal lower extremity strength, and regained ambulation with assistance before discharge from the hospital 8 days later. Subsequent outpatient follow-up visits showed progressive improvement in symptoms. At 2 weeks postoperatively, the patient recovered the ability to walk independently though still utilizing a rolling walker. At 6 weeks postoperatively, the patient had regained full strength and independent ambulation without assistive devices. At the 3-month follow-up, his only residual symptom was numbness in both feet, and he had returned to work with limitations only on heavy lifting. At 1 year, he continued to work full-time without limitations, reporting mild back pain and surgical site tightness but denied any sensory loss, motor weakness, or issues of incontinence. At 1 year, CT and radiography of the spine showed stable postoperative bony fusion from T5 to T12 with stable instrumentation and dense sclerosis involving the T7–10 vertebral bodies (Figs. 4B and 5B). A radiograph of the spine at 2 years showed stability as compared to the 1-year results (Fig. 4C). Beyond occasional mild back pain, surgical site tightness, and occasional thigh paresthesias, the patient reported no other complaints in the 2-year postoperative period.

**Patient Informed Consent**

The necessary patient informed consent was obtained in this study.

**Discussion**

**Observations**

The diagnosis of PDB is clinical and supported by laboratory and imaging findings. Radiographs are the modality of choice, and findings can be as follows: “V-shaped” cortical medullary lesions in the lytic phase; cortical thickening with trabecular coarsening in the mixed phase; and diffuse bone sclerosis with cortical thickening in

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**FIG. 1.** Preoperative imaging. **A**: CT scan showing significant blastic and sclerotic lesions of the spine at levels T8–11. **B**: T1-weighted MRI consistent with CT findings and showing spinal canal stenosis at the T8–11 levels. **C**: T2-weighted MRI showing densely sclerotic lesions of the spine. **D**: Three-dimensional reconstruction of CT images.
the sclerotic phase. CT scans allow better visualization of PDB’s mixture of cortical and trabecular bone and help differentiate PDB from blastic bony malignancies that often manifest vertebral sclerosis (“ivory vertebra”). Bone scintigraphy helps to stage the extent of the disease.

Bone hypervascularity is seen in active PDB, resulting from the replacement of the fatty marrow with fibrovascular tissue. This can lead to blood diversion and neural ischemia, causing myelopathy through a combination of direct compression and vascular insufficiency. On MRI, heterogeneous signals on T1 and T2 sequences suggest tissue replacement, with the postcontrast phase showing greater enhancement compared to unaffected bone, suggesting hyperemia. MRI is also helpful in assessing complications and preoperative planning.

PDB involving the spine is often asymptomatic, and when symptomatic, can be managed medically. Classically, the role of surgery in PDB has been limited to the removal of osteosarcoma, stabilization of fractures, and, in rare cases, symptomatic relief. This case study supports the latter, specifically in the setting of worsening myelopathy.

In this case, postsurgical CT imaging showed stable changes related to the laminectomies and fusion performed from T5 through T12 with no hardware complications. Interestingly, radiographic data at 1 year postoperatively indicated the presence of densely sclerotic bone involving the affected thoracic vertebrae (T7–10), reflecting the characteristic bone remodeling associated with Paget's disease after bisphosphonate treatment. This increased sclerosis and density observed postsurgery are indicative of the disease’s dynamic nature and the body’s response to medical and surgical intervention. Both FIG. 2. Axial T2-weighted MRI at T9 demonstrating severe stenosis with complete obliteration of surrounding cerebrospinal fluid. Circumferential disease is evident, with the most pronounced bony disease posteriorly.

FIG. 3. Technetium-99m bone scans showing diffusely increased activity in the lower thoracic spine, consistent with PDB.
FIG. 4. A: CT scan obtained immediately following implantation showing stable fixation of the thoracic spine. B: One-year CT scan showing sclerotic activity with mild scoliosis. C: A 1.5-year CT scan showing stable changes in the thoracic spine with intact hardware.

FIG. 5. A: Radiograph obtained immediately following implantation showing stable fixation of the thoracic spine. B: One-year radiograph showing sclerotic activity with mild scoliosis. C: Two-year radiograph showing stable changes of the thoracic spine with intact hardware.
Lessons

Though a rare diagnosis, PDB involving the spine should be considered in the differential of bony lesions of the spinal column. PDB has a stereotypical radiographic appearance with a mixture of cortical and trabecular bone and is characterized by elevations in markers of bone turnover. First-line management of PDB of the spine should be the administration of intravenous bisphosphonates unless defined operable conditions exist such as superimposed malignant tumors, unstable fractures, or severe refractory myelopathy. Calcitonin and denosumab may be added to the bisphosphonate regimen for difficulties managing neurological pain or associated giant cell tumors, respectively. Initiation of medical treatment has been associated with a significant reduction of bone pain and complete reversal of clinical symptoms of spinal stenosis including myelopathy. If the medical treatment fails to reach pain goals, or if previously identified conditions exist, surgical intervention is recommended. While data and guidelines for surgical intervention in PDB are limited, surgical decompression in patients with myelopathy is often effective. A systematic review conducted in 2016 led by Jorge-Mora et al. indicated that significant symptomatic benefits could be observed in patients undergoing spine surgery for PDB. In this study, 17 patients across 9 studies underwent varying combinations of laminectomies, vertebroplasties, and fusions for correction of the chief complaint of pain (2, 12%) or neurological deficits (15, 88%). Of the 17 patients, 14 saw improvements at 1 month postoperatively, 2 saw improvements in neurological symptoms past the 1-month follow-up, and 1 died 2 weeks later from cardiorespiratory failure. Open surgeries were associated with a complication rate of 25%, with the most common being significant bleeding. Antipagetic therapy, such as bisphosphonates, before the intervention has been shown to reduce intraoperative blood loss and the risk of revision surgery and should be considered preoperatively in all patients.

Despite the effectiveness of surgical treatment, given the high rate of complications, the decision for upfront surgical treatment versus medical management of myelopathy secondary to PDB remains undecided, as medical therapy alone may provide effective treatment. Multiple examples of the effectiveness of medical therapy are documented. Feki et al. reported a 62-year-old patient with both thoracic and lumbar spinal cord compression secondary to PDB, treated with an intravenous infusion of 5 mg of zoledronic acid without surgical intervention. The patient saw a decrease in ALP as well as objective improvements in power and sensory deficits. Future studies focused on developing a rational treatment model for compressive spinal PDB with an emphasis on surgical versus medication intervention may be helpful.

In addition to initial management, postsurgical monitoring and medical management of PDB patients are critical, as the progression of osteolytic activity may occur locally or a patient may develop other areas of PDB following surgery. Rolvien et al. presented a male patient who had undergone 7 spinal operations related to lumbar PDB over 12 years due to disease progression with observation or only oral bisphosphonates. These authors went further to say that regular rechecking of bone turnover and the administration of postoperative intravenous zoledronate are necessary to prevent the recurrence of pain. PDB is often characterized by osteolytic then progressive osteoblastic activity and finally an osteoquiescent phase, which may predispose postoperative patients to hardware failures. We concur that regular postoperative monitoring of bone-turnover markers and radiographs is critical to identify disease progression and confirm implant stability. Our patient’s postsurgical CT imaging showed notable sclerosis in the affected vertebrae and stabilization of markers of bone turnover, highlighting the dynamic nature of bone remodeling in PDB after medical treatment.

On rare occasions, PDB can promptly transition from asymptomatic to significant impairment and necessitate surgical intervention. This case supports the growing evidence that surgical intervention, particularly decompressive surgery, can provide symptomatic relief in cases of myelopathy secondary to PDB.
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