

# Spinal Myeloid Sarcoma “Chloroma” Presenting as Cervical Radiculopathy: Case Report

Xiaobang Hu<sup>1</sup> Imran Shahab<sup>2</sup> Isador H. Lieberman<sup>1</sup>

<sup>1</sup>Scoliosis and Spine Tumor Center, Texas Back Institute, Texas Health Presbyterian Hospital Plano, Plano, Texas, United States

<sup>2</sup>MD Pathology, Plano, Texas, United States

Address for correspondence Isador H. Lieberman, MD, MBA, FRCSC, Scoliosis and Spine Tumor Center, Texas Back Institute, Texas Health Presbyterian Hospital Plano, 6020 W. Parker Road, Suite 200a, Plano, TX 75093, United States (e-mail: ilieberman@texasback.com).

Global Spine J 2015;5:241–246.

## Abstract

**Study Design** Case report.

**Objective** Myeloid sarcoma (also known as *chloroma*) is a rare, extramedullary tumor composed of immature granulocytic cells. It may occur early in the course of acute or chronic leukemia or myeloproliferative disorders. Spinal cord invasion by myeloid sarcoma is rare. The authors report a rare case of spinal myeloid sarcoma presenting as cervical radiculopathy.

**Methods** A previously healthy 43-year-old man presented with progressive neck, right shoulder, and arm pain. Cervical magnetic resonance imaging (MRI) revealed a very large enhancing extradural soft tissue mass extending from C7 through T1, with severe narrowing of the thecal sac at the T1 level. The patient underwent posterior cervical open biopsy, laminectomy, and decompression. Histologic examination of the surgical specimen confirmed the diagnosis of myeloid sarcoma. Postoperatively, a bone marrow biopsy was done, which showed myeloproliferative neoplasm with eosinophilia. The patient then received systemic chemotherapy and radiotherapy.

**Results** At the 10-month follow-up, the patient reported complete relief of arm pain and neck pain. X-rays showed that the overall cervical alignment was intact and there was no evidence of a recurrent lesion. MRI showed no evidence of compressive or remnant lesion.

**Conclusions** Spinal myeloid sarcoma presenting as cervical radiculopathy is rare, and it may be easily misdiagnosed. Knowledge of its clinical presentation, imaging, and histologic characterization can lead to early diagnosis and appropriate treatment.

## Keywords

- ▶ myeloid sarcoma
- ▶ chloroma
- ▶ spine
- ▶ cervical radiculopathy

## Introduction

Myeloid sarcoma (also known as *granulocytic sarcoma* or *chloroma*) is a rare, extramedullary tumor of immature myeloid cells. It more often develops in the setting of acute myeloid leukemia (AML), but it can also occur in association with chronic myeloid leukemia, myelodysplastic syndrome, or, rarely, in the absence of marrow involvement.<sup>1</sup> The clinical presentation of myeloid sarcomas varies, and it is largely dependent on the site of involvement. The most common locations include the skin, soft tissue, bone, periosteum, and lymph nodes.<sup>1</sup> Spinal epidural myeloid sarcoma is uncom-

mon, and spinal cord compression caused by a myeloid sarcoma is exceptionally rare.

Here we report an unusual case of cervical radiculopathy as the first presentation of a spinal epidural myeloid sarcoma. The patient was successfully treated by spinal decompression and subsequent chemotherapy and radiation.

## Case Report

### History and Examination

A 43-year-old man presented with progressive neck, right shoulder, and arm pain of about 2 weeks' duration. The patient

received  
August 1, 2014  
accepted  
February 10, 2015

DOI <http://dx.doi.org/10.1055/s-0035-1549433>.  
ISSN 2192-5682.

© 2015 Georg Thieme Verlag KG  
Stuttgart · New York

License terms

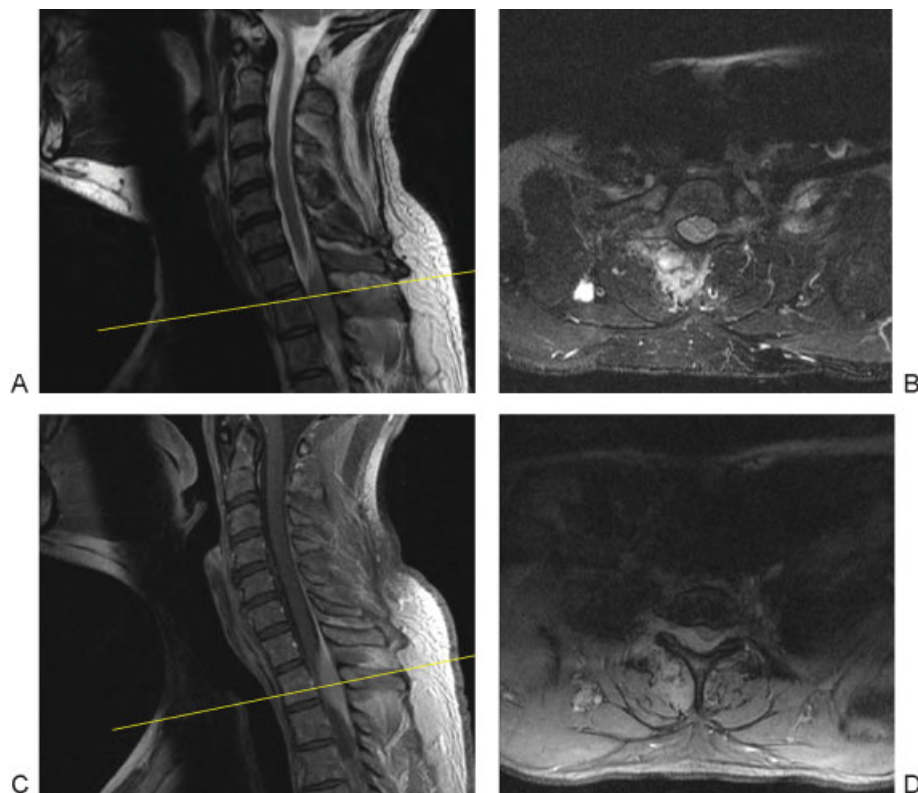




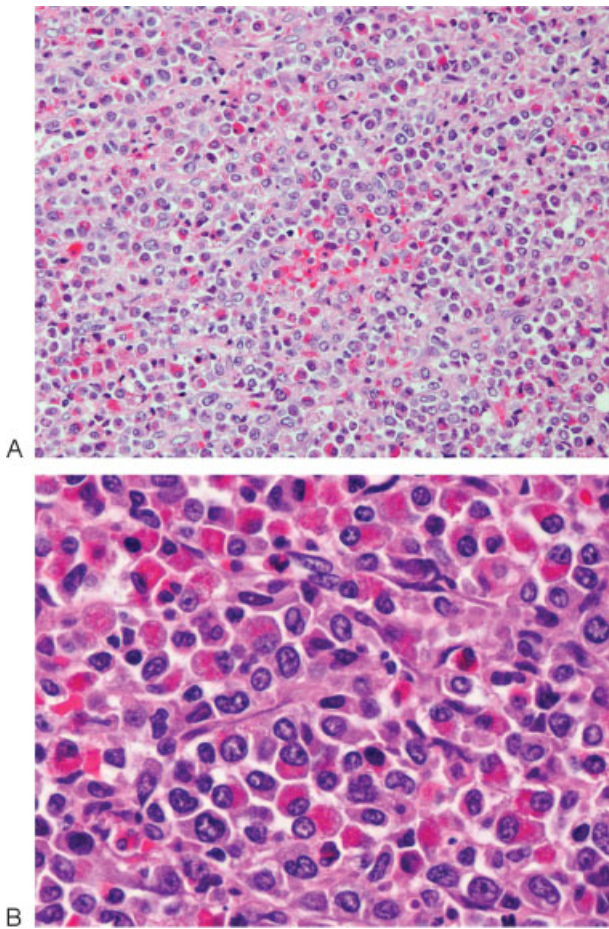
**Fig. 1** (A–C) Anteroposterior and lateral extension and flexion X-rays showed mild multilevel cervical degenerative changes with spurring noted at C5–C7. Disk height was relatively well maintained.

described that the pain radiated down his right upper extremity through the triceps region, into the forearm, and onto the fourth and fifth digits. There was no precipitating factor such as injury or trauma. The patient initially sought care with a chiropractor and a nurse practitioner and received various treatments including massage, cervical adjustment, intramuscular steroid injection, and steroid pack. He was also prescribed muscle relaxants and narcotic analgesics. Despite these

interventions, his pain continued to increase. On physical examination, the patient was noted to be splinting his neck and actively resisting any neck motion. There was tenderness palpable over the right scapular border. There was decreased sensation on the ulnar aspect of his right hand as well as decreased pinch strength and sensation in the right fourth and fifth digits. There was also decreased sensation to pinprick over the entire right scapula. X-ray examination showed mild,



**Fig. 2** Cervical magnetic resonance imaging showing the enhancing extradural soft tissue mass extending from C7 through T1 with severe narrowing of the thecal sac at T1 level. (A, B) T2-weighted images; (C, D) T1-weighted images.

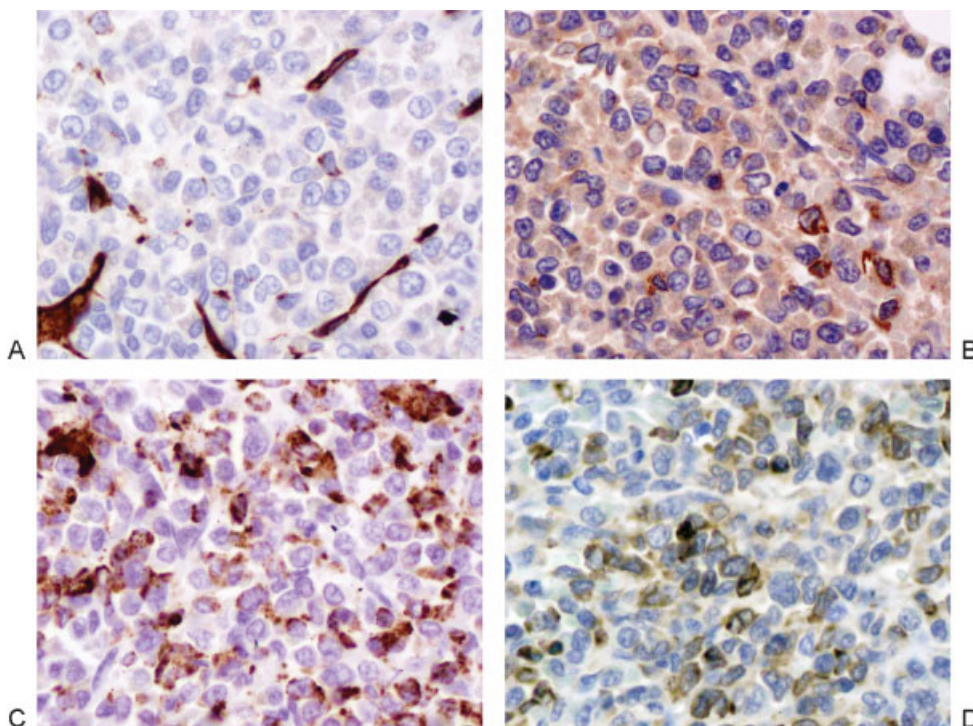


**Fig. 3** (A, B) Histologic examination showed extensive involvement by a hematopoietic neoplasm composed of atypical cells with irregular nuclei and delicate chromatin. Many immature eosinophils were noted accompanying the neoplasm.

multilevel cervical degenerative changes with spurring noted at the C5, C6, and C7 levels, but the disk height was relatively well maintained (► **Fig. 1**). Cervical magnetic resonance imaging (MRI) revealed a very large, enhancing extradural soft tissue mass extending from C7 through T1 with severe narrowing of the thecal sac at the T1 level. There was also a large soft tissue mass to the right of midline from C7 to T2 with some areas of decreased or absent enhancement suggesting necrosis or developing phlegmon or abscess (► **Fig. 2**).

**Operation**

In view of the patient’s progressive neurologic symptoms and signs, combined with the MRI findings, we elected to decompress the spine and obtain an open definitive biopsy through a tumor-type approach. The risks, benefits, and alternatives to various diagnostic and therapeutic measures, including the merits of a needle biopsy, were initially discussed with the patient, and considering that the needle biopsy itself carries risk and the likelihood of a false-negative biopsy was quite high, we recommended and he decided to proceed with the definitive procedure. The patient then underwent a posterior cervical open biopsy, laminectomy, and tumor decompression. The large epidural mass indenting the thecal sac was totally removed during the surgery, and there were no intraoperative complications. Histologic examination showed extensive involvement by a hematopoietic neoplasm composed of atypical cells with irregular nuclei and delicate chromatin. Many immature eosinophils were noted, accompanying the neoplasm. The neoplastic cells infiltrated skeletal muscle fibers, fibrous tissue, and fat. A few mitotic figures were present. A few neoplastic cells showed indented nuclear membranes (► **Fig. 3**). Immunohistochemical stains showed extensive positive staining in the atypical cells with CD43, as



**Fig. 4** Immunohistochemical stains showed positive staining in the atypical cells with CD34 (A), CD117 (B), CD68 (C), and lysozyme (D).



**Table 1** Patient-reported outcome at the latest follow-up

	Preoperative	10 mo postoperative
Arm pain score (VAS)	8	0
Neck pain score (VAS)	4	0
NDI score	17	0

Abbreviations: NDI, Neck Disability Index; VAS, visual analog scale.

well as focal staining in these cells with CD10, CD34, CD 68, CD117, and lysozyme also noted (►Fig. 4). CD3 and CD20 stains showed a few reactive T and B cells. S-100 stain showed nonspecific granular staining in the immature eosinophils. No staining with CD30 was noted. Cytogenetic fluorescence in situ hybridization analysis performed on the paraffin-embedded sections from the paraspinal mass was positive for deletion of the *CHIC2* gene, indicating fusion of *FIP1L1/PDGFR*A. The morphologic and immunologic features led to the diagnosis of myeloid sarcoma.

### Postoperative Course

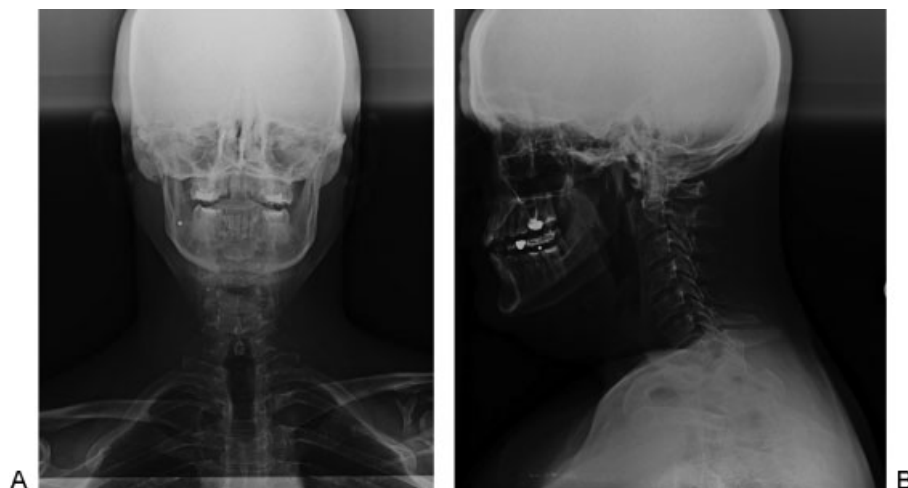
The patient was referred to an oncologist postoperatively, and a subsequent bone marrow aspiration verified features of a myeloproliferative neoplasm with eosinophilia. The bone marrow also showed *FIP1L1-PDGFR*A rearrangement. Fluorescence in situ hybridization analysis on the bone marrow aspirate was positive for deletion of the *CHIC2* gene in 28.5% of 200 interphase nuclei and was negative for *BCR/ABL1*, *PDGFRB* rearrangement, *FGFR1* rearrangement, *RUNX1T1/RUNX1*, and *MLL*. The patient then received chemotherapy followed by radiation therapy. At his 10-month follow-up visit, the patient reported no arm pain, no neck pain, and significantly improved Neck Disability Index scores (►Table 1). Physical examination showed 5 of 5 strength in his upper extremities. X-rays showed that the overall cervicothoracic junctional alignment was maintained, and there was no evidence of

any new lesions (►Fig. 5). MRI showed no evidence of compressive or remnant lesion (►Fig. 6).

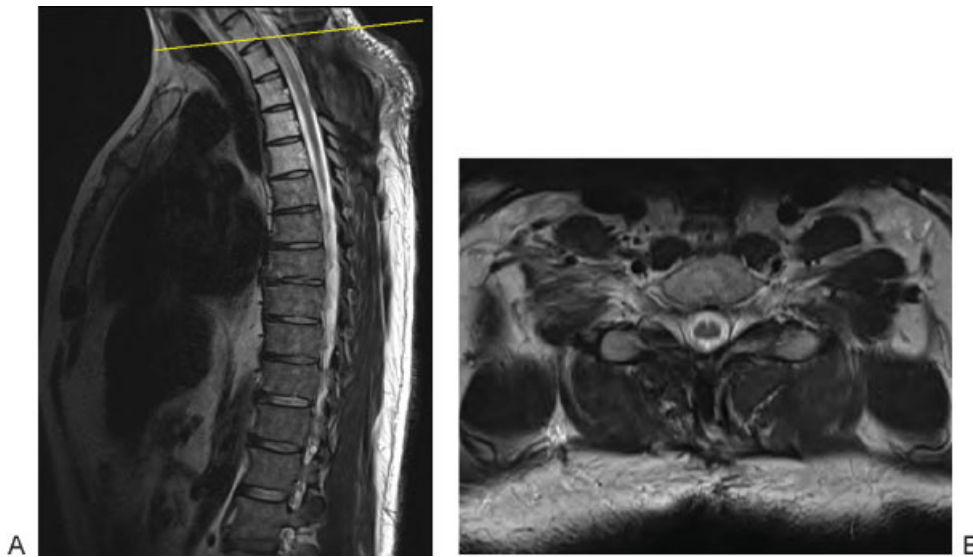
### Discussion

Myeloid sarcoma was first described in 1811 and later named *chloroma* by King because of the green color caused by the presence of myeloperoxidase granules in the malignant myeloid cells.<sup>2-5</sup> Overall, myeloid sarcoma represents a rare hematologic phenomenon with an incidence of 2.5 to 9.1% in patients with AML.<sup>1,6</sup> It could occur concomitantly, following, or, rarely, before the onset of systemic bone marrow leukemia.<sup>1,6,7</sup> The prevalence of myeloid sarcoma in the spine is estimated to be less than 1% among all patients with acute and chronic myeloid leukemia.<sup>5</sup> All levels of the spine can be affected, but it seems more frequent in the lumbosacral spine, followed by the thoracic spine and cervical spine.<sup>5</sup> The clinical manifestation of spinal myeloid sarcoma varies, and it largely depends on the tumor location and size. Spinal myeloid sarcoma that arises in the epidural or paraspinal tissues can present with signs of cord compression due to mass effect. In the presented case, the patient's initial presentation was neck, shoulder, and arm pain, and X-ray examination showed mild multilevel cervical degenerative changes. This unspecific presentation could easily be misdiagnosed with degenerative disk disease, which could lead to prolonged and inappropriate treatment and delay the timely diagnosis.

The diagnosis of spinal myeloid sarcoma is often challenging because the clinical and laboratory data are usually not contributory. The radiologic findings are usually nonspecific, and the mass often appears as a soft tissue mass on computed tomography or MRI.<sup>5,6</sup> On MRI examination, myeloid sarcoma of the spine has a similar signal intensity to muscle on T1-weighted images and has an intermediate signal intensity on T2-weighted images.<sup>5</sup> Immunohistochemistry is the most practical method for establishing the diagnosis of myeloid sarcoma, and a series of markers can be used for diagnosis confirmation and further lineage characterization.<sup>6</sup> If the



**Fig. 5** (A, B) At the latest follow-up (10 months postoperative), anteroposterior and lateral X-rays showed that the overall cervical alignment was intact.



**Fig. 6** (A, B) At the latest follow-up (10 months postoperative), magnetic resonance imaging showed no evidence of compressive or remnant lesion.

patient has established leukemia, the diagnosis of myeloid sarcoma is relatively straightforward, and it is suggested to always be included in the differential diagnosis for patients with AML who develop a soft tissue mass.<sup>6</sup> The diagnosis of myeloid sarcoma could be difficult if there is no clinical history of leukemia. In this setting, every effort should be made to obtain a tissue diagnosis.<sup>6</sup> In the present case, the patient underwent posterior cervical open biopsy, laminectomy, and tumor decompression, and immunohistochemical stains showed positive staining in the atypical cells with CD43, CD10, CD34, and CD117, which helped to confirm the diagnosis. Myeloid and lymphoid neoplasms with *FIP1L1-PDGFR*A fusion gene as shown in this case are extremely rare, and it is reported to have a striking, male predominance with features most commonly resembling chronic eosinophilic leukemia. It also rarely presents with concomitant AML, including myeloid sarcoma, and/or with concomitant T-cell lymphoblastic lymphoma.<sup>8,9</sup>

The treatment strategy for myeloid sarcoma largely depends on whether it develops at initial diagnosis or at relapse. The optimal timing and treatment of isolated myeloid sarcoma is still not clear; however, it has been reported that the patients who have delayed or inadequate systemic treatment of myeloid sarcoma will almost always progress to AML.<sup>9</sup> For the patient with myeloid sarcoma who presents with concurrent marrow involvement, systemic treatment directed at the underlying leukemia is always suggested. A combined approach, such as surgical resection, chemotherapy, radiotherapy, and bone marrow transplantation, has been emphasized.<sup>6,10</sup> Inoue et al<sup>11</sup> identified 26 patients without leukemia with myeloid sarcoma from the literature, and they found that the treatment strategy varied individually. Surgical decompression was performed in 22 patients, and 21 of the patients received additional AML-type systemic treatment. However, the patients' outcome was generally poor, especially in the patients who showed progression to leuke-

mia. Overall, prompt diagnosis and adequate treatment are essential to achieving a good outcome.<sup>6,12</sup>

## Conclusion

We report a rare case of spinal myeloid sarcoma presenting as cervical radiculopathy. This may be easily misdiagnosed. Knowledge of its clinical presentation, imaging, and histologic characterization can lead to early diagnosis and appropriate treatment.

## Disclosures

Xiaobang Hu, none  
Imran Shahab, none  
Isador H. Lieberman, none

## Ethical Board Approval

This study has been approved by Texas Health Resources Institutional Review Board (IRB) (study ID: Pro00004880).

## References

- Bakst R, Wolden S, Yahalom J. Radiation therapy for chloroma (granulocytic sarcoma). *Int J Radiat Oncol Biol Phys* 2012;82(5):1816–1822
- Burns A. *Observations of Surgical Anatomy in Head and Neck*. Edinburgh, Scotland: Thomas Royce; 1811
- King A. A case of chloroma. *Monthly J Med* 1853;17:97
- Reardon G, Moloney WC. Chloroma and related myeloblastic neoplasms. *Arch Intern Med* 1961;108:864–871
- Seok JH, Park J, Kim SK, Choi JE, Kim CC. Granulocytic sarcoma of the spine: MRI and clinical review. *AJR Am J Roentgenol* 2010;194(2):485–489
- Bakst RL, Tallman MS, Douer D, Yahalom J. How I treat extramedullary acute myeloid leukemia. *Blood* 2011;118(14):3785–3793

- 7 Kalayci M, Sümer M, Yenidünya S, Ozdolap S, Açıkgöz B. Spinal granulocytic sarcoma (chloroma) presenting as acute cord compression in a nonleukemic patient. *Neurol India* 2005;53(2):221–223
- 8 Chen D, Bachanova V, Ketterling RP, Begna KH, Hanson CA, Viswanatha DS. A case of nonleukemic myeloid sarcoma with FIP1L1-PDGFR $\alpha$  rearrangement: an unusual presentation of a rare disease. *Am J Surg Pathol* 2013;37(1):147–151
- 9 Klion AD. Eosinophilic myeloproliferative disorders. *Hematology (Am Soc Hematol Educ Program)* 2011;2011:257–263
- 10 Meis JM, Butler JJ, Osborne BM, Manning JT. Granulocytic sarcoma in nonleukemic patients. *Cancer* 1986;58(12):2697–2709
- 11 Inoue T, Takahashi T, Shimizu H, et al. Spinal granulocytic sarcoma manifesting as radiculopathy in a nonleukemic patient. *Neurol Med Chir (Tokyo)* 2008;48(3):131–136
- 12 Cui Y, Zhou JL, Wu JH, Zhang JZ. Synchronous granulocytic sarcoma of the breast and spine: a case report and review of the literature. *Chin Med J (Engl)* 2008;121(18):1854–1856

## Editorial Perspective

*EBSJ* thanks the authors of the original case submission and the commentary. The value of having a multidisciplinary and global readership is reflected in this case presentation and discussion. *EBSJ* wishes to make a technical and a more general commentary based upon these two submissions.

The vulnerability of the cervicothoracic junction toward developing a progressive kyphotic deformity following a laminectomy was pointed out by Dr. Delashaw and is a substantial concern that actually rises over time. Of course, it is desirable to have MRI available as neuroimaging modality to have a handle on disease recurrence and study cord signal changes. Hardware, even in form of titanium, will cause some signal interference, limiting the visualization of these critical areas. Computed tomography can be a helpful secondary imaging modality for extradural lesions but will be limited in looking at intradural or intramedullary pathologies. That said—if in doubt, supplemental posterior segmental instrumentation and arthrodesis of the cervicothoracic junction in the presence of decompressive laminectomies is probably preferable, as late deformity correction of a postlaminectomy

cervicothoracic kyphosis usually is a far more challenging and morbid undertaking.

The second comment pertains to the circuitous road to arriving at a diagnosis. Timely diagnosis of many spine-related diseases in the presence of comorbidities is not infrequently an arduous undertaking with many pitfalls, as seen in this case. In an era of increasing emphasis on “cookbook” medicine with mandated implementations of relatively simplistic symptom suppressive management algorithms, delays in diagnosis of more unusual but serious pathologies may become more of a norm rather than the exception. To date, formal clinical pathways in spine care struggle with the threat of missed early management opportunities with serious pathologies such as neoplasia. The very real risk of unreflective insistence of practicing “evidence-based medicine” in form of a “epidemiologic medicine” by relying on statistical incidences of a majority of diseases leaves the very real risk of having more rare but significant pathologies be pushed aside until patients present so late in their disease process that there is little to do but practice “damage control medicine.”