e-ISSN: 0975-1556, p-ISSN:2820-2643

Available online on www.ijpcr.com

International Journal of Pharmaceutical and Clinical Research 2024; 16(5); 494-500

Original Research Article

Compressive Thoracic Myelopathy: An Unexpected Adversary Met on Table

Gayatri Brahmandam[1], Kumar Lakshman[2], Abhishek Chowdhury[3], Deekshitha K Aiyer[4], Anita Mahadevan[5], Triza Kumar Lakshman[6]

[1]Junior Resident, Department of Neurosurgery, Adichunchanagiri Institute of Medical Sciences;
[2]Associate Professor, Department of Neurosurgery, Adichunchanagiri Institute of Medical Sciences,
[3]DM Neuropathology Resident, NIMHANS
[4]Junior Resident, Department of General Surgery, Adichunchanagiri Institute of Medical Sciences;
[5]Professor and Head, Department of Neuropathology, NIMHANS,
[6]Professor, Department of OBG, Adichunchanagiri Institute of Medical Sciences

Received: 25-02-2024 / Revised: 23-03-2024 / Accepted: 26-04-2024
Corresponding Author: Dr. Kumar Lakshman
Conflict of interest: Nil

Abstract:

Thoracic myelopathy is a relatively overlooked cause of spinal disorders with possible diagnoses including pathological fractures, Pott's disease, Multiple Myeloma and non-Hodgkin's Lymphoma. In India, symptoms resembling Pott's Spine necessitate prompt and effective action as it is the most common extranodal manifestation of tuberculosis. Additionally, it can resemble the aforementioned diseases on radiological examination leading to complications and oversight in patient care. Therefore, a thorough workup is indicated when a patient presents with symptoms of myelopathy. This case report will provide a succinct overview of the presentation and novel management of a patient with a high-grade non-Hodgkin's Lymphoma – B Cell Subtype with investigations suggesting Pott's disease.

Keywords: Non-Hodgkins lymphoma, PSEL, Potts Spine.

Introduction

Spinal cord compression can be traumatic or atraumatic. While traumatic causes usually require emergency care and immediate stabilisation, atraumatic causes such as degenerative spondylosis with myelopathy, metastatic disease of the spine, primary spinal cord malignancy and spinal epidural abscess may have an insidious course giving clinicians more time to diagnose and manage such patients. [1]

The cervical and lumbar spine is more commonly involved in myelopathy and other causes of spinal cord compression. Thoracic myelopathy is a relatively overlooked cause of atraumatic spinal disorders with possible diagnoses including pathological fractures, Pott's disease, Multiple Myeloma and non-Hodgkin's Lymphoma. [2] In India, symptoms suggestive of Pott's Spine necessitate prompt and effective action as it is the most common extranodal manifestation of tuberculosis. Additionally, it can resemble the aforementioned diseases on MRI leading to complications and oversight in patient care. [3] There is extensive literature documenting the presentation, management and follow-up of patients with Pott's Spine. On the flip side, literature about Primary Spinal Epidural Lymphoma (PSEL) is relatively scarce, especially in the management as the treatment options are ambiguous. [4] It is generally regarded that surgery and radiotherapy provide good patient outcomes as B cell Lymphoma is responsive to radiation therapy, chemotherapy and surgical stabilisation.

Also to be considered here is the incidence of PSEL. It only accounts for 0.9% of extranodal non-Hodgkin's Lymphoma. [5] Primary Diffuse Large B Cell Lymphoma has an incidence of 2 in 10 million. The presenting symptoms of myelopathy usually include low back pain, weakness in the lower limbs, an inability to walk and, in the later stages, bowel and bladder irregularities. Here, we present a patient with thoracic compressive myelopathy who was managed with surgical stabilisation and chemotherapy.

Case:

A 65-year-old female presented to our hospital with complaints of low back pain for one month and new onset bilateral leg weakness since 5 days. This weakness was associated with an inability to walk and a tingling sensation in the lower limbs. There

was no history of fever, cough or night sweats. The patient denied a history of a fall or any other trauma. She denied any bowel or bladder disturbances. Her medical history is significant for hypertension for which she has been on antihypertensives for a year now. She has no other chronic illnesses.

On examination, her vitals excluding blood pressure were within normal limits. Her blood pressure was elevated, attributed to her hypertension. Systemic examination was unremarkable. CNS motor examination showed a power of 4/5 in both lower limbs. It was assessed by Medical Research Council grading. Additionally, Babinski's sign was equivocal on both sides and reflexes were brisk with a score of +2.

Sensory examination revealed paraesthesia below the subcostal level with an associated tingling sensation. Her vitals were stable and breast and thyroid examination revealed no abnormalities. Contrast-enhanced MRI of the dorsal and lumbosacral spine was done and it showed D5 spondylodiscitis with homogenously enhancing epidural, pre and paravertebral soft tissue components with resultant significant spinal canal stenosis and cord compression. This was suggestive of Pott's Spine. There was also an epidural soft tissue component seen encasing the thecal sac. CECT showed a lytic lesion involving posterior elements of the D5 vertebra with an associated small soft tissue density lesion on the right side. Although there was initially a suspicion of Azygous vein involvement, it was ruled out with the CECT Thorax.

Investigations

Biochemical parameters at the time of admission are recorded below.

Table 1:

Lab Parameters	**Values (Units)**
Haemoglobin	10.1 gm/dL
WBC Count	14500 cells/cumm
Platelet count	3.10 lakhs/cumm
Creatinine	0.6 mg/dL
AST	61 U/L
ALT	49 U/L
Alkaline Phosphatase	156 U/L
Total Bilirubin	1.6 mg/dL
HIV testing	Negative
Bleeding Time	2 minutes 15 seconds
Clotting Time	5 minutes 45 seconds
Random Blood Sugar	128 mg/dL

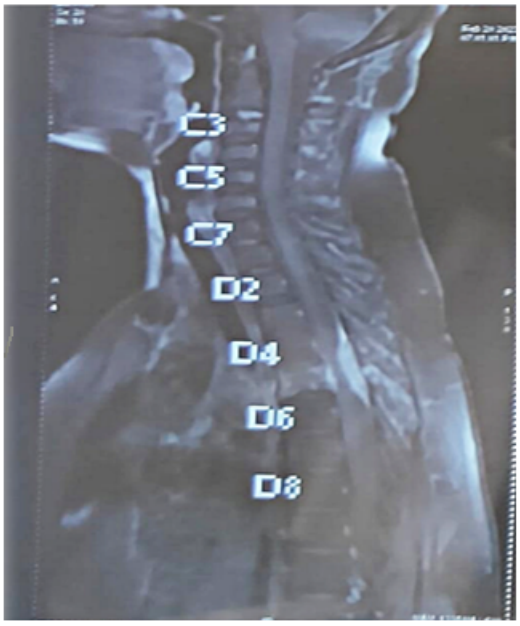

Figure 1: A labelled CE MRI frame sagittal section showing homogenously enhancing soft tissue component at D5 level

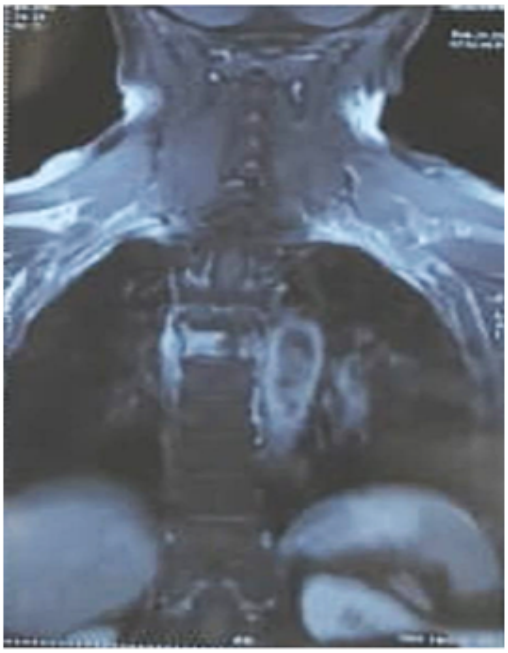

Figure 2: Paravertebral involvement at D5 vertebral level in coronal section

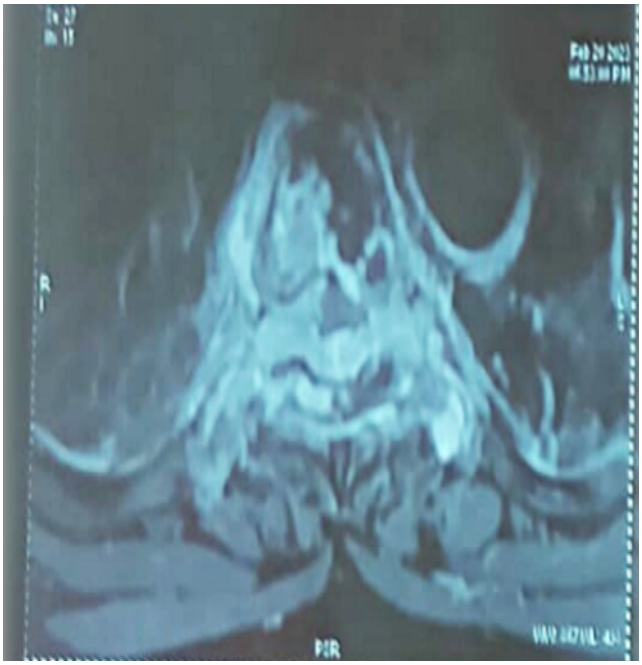

Figure 3: Axial section showing increased uptake of contrast in the epidural, pre and paravertebral region at D5 vertebral level

She was started on empirical anti-tubercular therapy and 48 hours later, underwent D4-D6 spinal fusion with implant placement. Posterior stabilisation was done with the evacuation of the epidural soft tissue component. The excised specimen was sent for biopsy as well as CBNAAT.

Intra OP Images

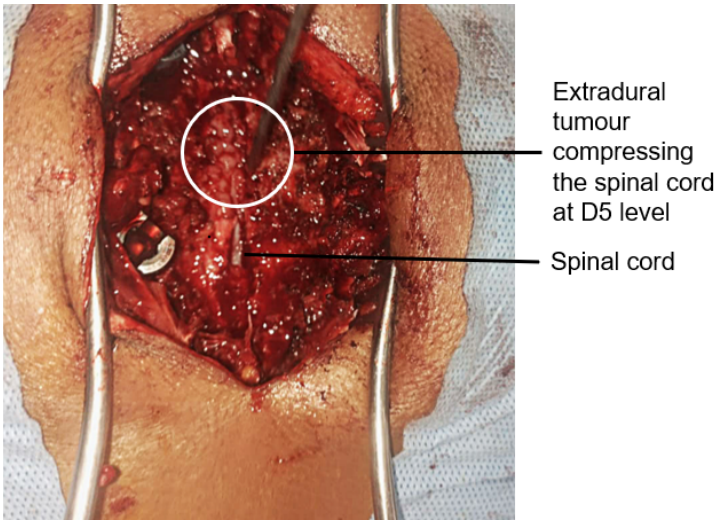

Figure 4: Tumour compressing the spinal cord at D5 level

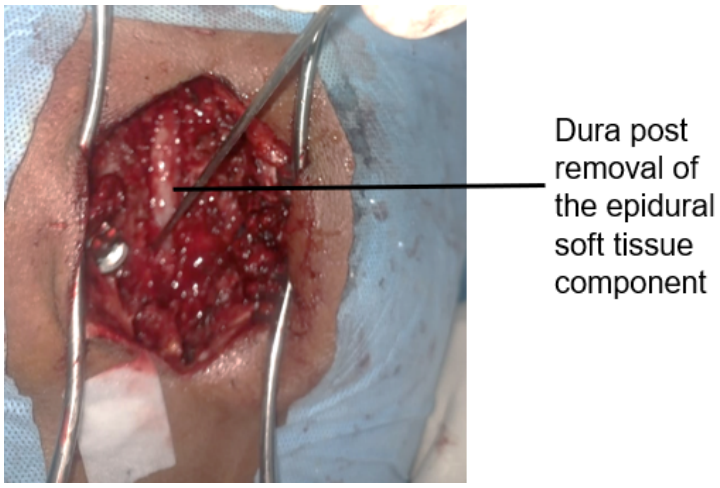

Figure 5: Dura post removal of the epidural soft tissue component

Post Operative Imaging

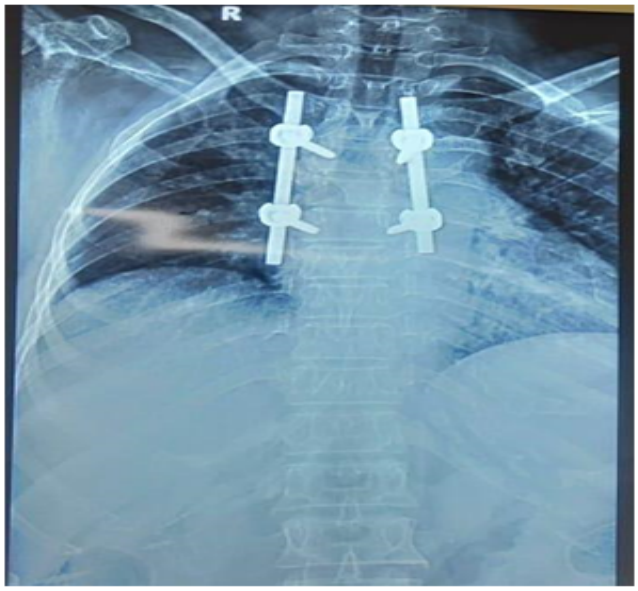

Figure 6: Post operative Radiograph in posteroanterior view

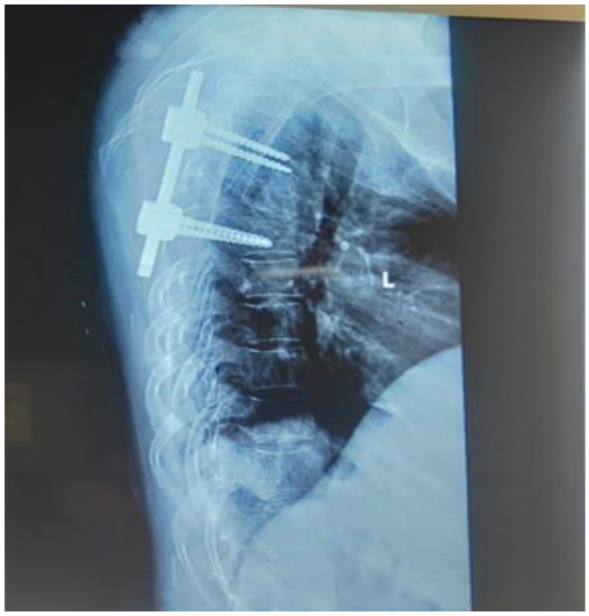

Figure 7: Postoperative radiograph in lateral view

The patient's recovery post-surgery was unremarkable.

Histopathology Report:

The biopsy report uncovered that the specimen was malignant, showing fragments of bony trabecular with marrow spaces and adjacent fibro adipose tissue and ligament containing dense infiltrates of CD20-positive large lymphoid cells showing features of dysplasia. The final diagnosis was that of a high-grade non-Hodgkins Lymphoma - B Cell type at the D5 vertebral level.

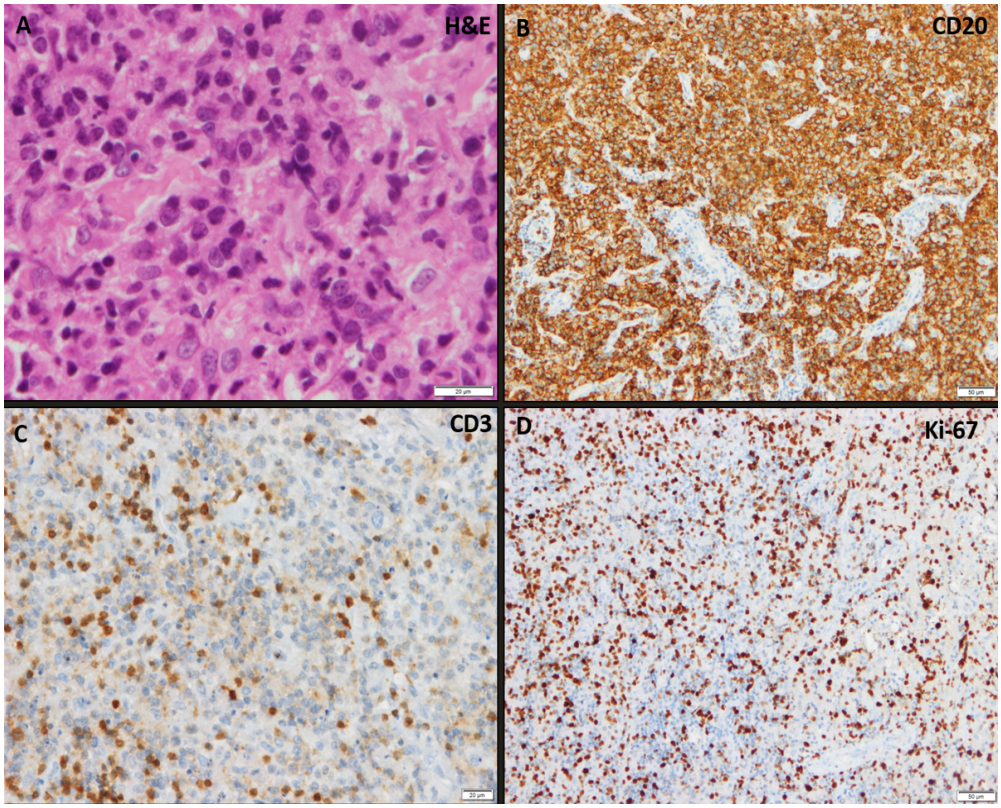

Figure 8: Microphotographs show characteristic features of high-grade non-Hodgkin's lymphoma with neoplastic lymphoid cells (A), that are of B cell phenotype (CD20+, B). Admixed mature reactive T cells (CD3+, C) are seen. The tumour cells exhibit very high Ki67 (MIB-1) labelling index (D). [A: H&E; B-D: immunoperoxidase. Magnification=scale bar]

Further Course:

The patient was referred to the oncology wing wherein she underwent a PET Scan which reported no sites of metastasis. There was increased uptake at the site of surgery, which indicated post-operative inflammation.

Following this, she was started on the R CHOP regimen which consists of the drugs Rituximab - a CD20 inhibitor, Cyclophosphamide - an alkylating agent, Doxorubicin - a topoisomerase II inhibitor, Vincristine - a chemotherapeutic agent that acts by inhibiting intracellular microtubule formation and finally, Prednisone - a steroid.

This is the most widely followed treatment for Diffuse Large B Cell Lymphoma.

Discussion:

By definition, Primary Spinal Epidural Lymphomas (PSEL) are tumours with a characteristic histopathological picture of lymphoma which are seen purely in the spinal epidural space, with an accompanying negative diagnostic workup for lymphoma at other sites. [7]

PSEL accounts for close to 10% of the cases of spinal lymphomas but the epidural space is a rare location. [8] At the time of presentation, which happens in two phases according to Epelbaum et al i.e., it may be in the prodromal phase, wherein the patient presents with localized back pain and occasional radicular pain and a second rapid neurological deterioration phase which usually occurs over 2 to 8 weeks. [9] Pott's Spine also can present with similar complaints at this stage making for an intriguing diagnostic puzzle. Additionally, "B" symptoms such as fever, night sweats and Pel Ebstein fever may be seen at the time of presentation with PSEL.

Primary Spinal Epidural Lymphoma is a potentially curable disease provided surgery is done and a multifaceted approach is adopted at the time of diagnosis. [7] The constellation of symptoms seen in PSEL overlaps with those of any epidural tumour, namely back pain at the level of the tumour, weakness of limbs and sensory deficits below the involved level and impairment of bowel and bladder at the later stages. [8] Imaging alone cannot distinguish between the various solid etiologies of compressive myelopathy.

In addition to being the most common primary spinal lymphoma, non-Hodgkins lymphoma most often involves the thoracic spine. This preponderance is attributed to its plasticity, which enables the development of bulky disease in the region. [10] The investigations that make up the diagnostic workup are X-ray spine or CT where the absence of bone destruction and erosion suggests the diagnosis of spinal lymphoma. On MRI, the mass presents as an iso-intense or hypo-intense homogenous tumour involving multiple segments and possible foraminal extension. [11,12]

Although not a consistent finding, the absence of hyperintensity on MRI may help differentiate it from hematomas and metastasis. Needle biopsy, despite being helpful for early diagnosis, does not have a satisfying diagnostic rate. [13]

No tried and tested treatment algorithm exists to treat spinal epidural lymphomas due to their low incidence. A multimodal approach is adopted which includes chemotherapy, surgical stabilisation, radiation therapy, steroids and stem cell transplant. Two chemotherapy regimens are followed currently - Hyper-CVAD and CHOP. [14] High-dose methotrexate (3.5g/m2) can be added along with Leucovorin rescue. Xiong L. et al calculated the Overall Survival (OS) and Disease-Free Survival (DFS) using the Kaplan Meier method and log-rank tests placing the 3-year OS at 81.1% and DFS at 46.3%. [8,15] The sample size was 130 including patients and case reports. [8]

It is worth noting that the RCHOP regimen has been documented to cause reduced Vertebral Density, even two years post-treatment, according to a study conducted by Svendsen P, et al. [16]

In conclusion, Compressive Thoracic Myelopathy is a relatively uncommon occurrence and Pott's Spine accounts for most cases of it. Therefore, a clinician should have a high index of suspicion for other causes. It is pivotal to keep in mind other diagnoses such as PSEL which is a diagnosis of exclusion, rather than inclusion.

References:

1. Singleton JM, Hefner M. Spinal Cord Compression. StatPearls Publishing; 2023
2. Ando K, Imagama S, Kobayashi K, Ito K, Tsushima M, Morozumi M, et al. Clinical features of thoracic myelopathy: A single-center study: A single-center study. J Am Acad Orthop Surg Glob Res Rev [Internet]. 2019; 3(11): e18.00090.
3. Prapruttam D, Hedgire SS, Mani SE, Chandramohan A, Shyamkumar NK, Harisinghani M. Tuberculosis--the great mimicker. Semin Ultrasound CT MR [Internet]. 2014; 35(3):195–214.
4. Bhardwaj P V, Abraham A, Alluri S (September 08, 2022) Primary Spinal Epidural Diffuse Large B-cell Lymphoma: Case Report and Literature Review. Cureus 14(9): e28934.
5. Pokhrel NB, Prasad R, Paudel S, Kafle D, Pokharel RK. Primary spinal epidural non-Hodgkin's diffuse large B-cell lymphoma: A case report. Clin Case Rep. 2020 Sep 21; 8(11):2276-2280.

6. Ansari S, Amanullah MF, Ahmad K, Rauniyar RK. Pott's spine: Diagnostic imaging modalities and technology advancements. N Am J Med Sci [Internet]. 2013; 5(7):404–11.
7. Cugati G, Singh M, Pande A, Ramamurthi R, Balasubramanyam M, Sethi S, et al. Primary spinal epidural lymphomas. J Craniovertebr Junction Spine [Internet]. 2011; 2(1):3.
8. Xiong L, Liao L-M, Ding J-W, Zhang Z-L, Liu A-W, Huang L. Clinicopathologic characteristics and prognostic factors for primary spinal epidural lymphoma: report on 36 Chinese patients and review of the literature. BMC Cancer [Internet]. 2017; 17(1).
9. Epelbaum R, Haim N, Ben-Shahar M, Ben-Arie Y, Feinsod M, Cohen Y. Non-Hodgkin's lymphoma presenting with spinal epidural involvement. Cancer [Internet]. 1986; 58(9): 2120–4.
10. Portenoy RK. Evaluation of back pain caused by epidural neoplasm. J Back Musculoskelet Rehabil [Internet]. 1993; 3(2):44–52.
11. Mullins GM. Malignant lymphoma of the spinal epidural space. Ann Intern Med [Internet]. 1971; 74(3):416.
12. Haque S, Law M, Abrey LE, Young RJ. Imaging of lymphoma of the central nervous system, spine, and orbit. Radiol Clin North Am [Internet]. 2008; 46(2):339–61.
13. Mascalchi M, Torselli P, Falaschi F, Dal Pozzo G. MRI of spinal epidural lymphoma. Neuroradiology [Internet]. 1995; 37(4):303–7.
14. Reed A, Sommerhalder D. The use of R-hyper-CVAD in a rare case of primary bone marrow diffuses large B-cell lymphoma. J Hematol (Brossard) [Internet]. 2019; 8(4):165–7.
15. Tang Y, Yang X, Xiao J, Liu K, Yan W, Song D, et al. Clinical outcomes of treatment for spinal cord compression due to primary non-Hodgkin lymphoma. Spine J [Internet]. 2013; 13(6):641–50.
16. Svendsen P, Shekhrajka N, Nielsen KL, Vestergaard P, Poulsen MØ, Vistisen AK, et al. R-CHOP(-like) treatment of diffuse large B-cell lymphoma significantly reduces CT-assessed vertebral bone density: a single center study of 111 patients. Leuk Lymphoma [Internet]. 2017; 58(5):1105–13.