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CASE REPORT

Giant Chordoma of the Thoracolumbar Spine

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ABSTRACT

Chordomas are rare tumors from primitive notochord. They are usually located at sacrococcygeal region when it affects the spine. We present a case of giant chordoma in the thoracolumbar spine in Peru. A 43-year-old woman came to the hospital in Huanuco due to back pain and sensorymotor alterations in the lower limbs. Imaging confirmed an unresectable tumor involving the spine. Two years later, she came back with paraplegia, weight loss, severe anemia and complicated urinary tract infection. Magnetic resonance imaging confirmed a mass measuring 15×15×14 cm in T11, T12 and L1 which histopathology was consistent with chordoma. Chordoma is a rare tumor which should be included in the differential diagnosis of spinal tumors. An earlier diagnosis leads to more options of treatments and longer survival.

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Introduction

Chordomas are rare malignant neoplasms that arise from primitive notochordial remnants along the axial skeleton. Its incidence is lower than 1 in 1,000,000 worldwide. Its incidence is higher in men and elderly people.[1, 2] Fifty percent of the cases are found in the sacrococcygeal region and 32.8% are found in the mobile parts of the spinal column.[2, 3] Although these percentages are controversial in different studies.[1] Chordomas represent about 2-4% of primary bone neoplasms.[4]

Chordomas have a slow growth, with local destruction of the bone and extension into the adjacent soft tissues.[2, 5] They have an indolent course with multiple local recurrences and a survival that varies from four to twenty years.[6] Patients with tumors located along the lumbar vertebrae may present with pain, bladder dysfunction, or weakness of the lower extremities.[7]

The time to diagnosis averages 19 months.[8] The primary treatment is extensive surgical resection with well-defined margins; however, surgery is often difficult because of the tumor location. Adjuvant radiotherapy is also commonly performed, even though they are relatively resistant to the irradiation.[9, 10]

We present a female patient with giant chordoma who did not have clinical symptoms that accompanies the development and growth of the neoplasia in early stages, so that the patient came to the hospital in an advanced stage which limited the treatment options.

Case Presentation

A 43-year-old woman from Huanuco, Peru, without any major medical or surgical history, presented with a four-month history of pain in the thoracolumbar region, followed by decreased lower limb sensorymotor abilities and repeated urinary tract infections. Magnetic resonance imaging (MRI) revealed an expansive tumoral mass along the T8-L1 vertebrae with severe spinal canal stenosis from T9-T12. The lesion size was 13.4×11×12.9

cm. chondrosarcoma was considered as differential diagnosis, since it had expanded into the paravertebral space and had infiltrated the adjacent muscles. Contact with the patient was lost for two years. Two years later, a contrast enhanced computed tomography in axial, sagittal, and coronal sections of the spinal column was performed which showed an extensive non-enhancing hypodense expansive lesion with regular margins and a cystic component with calcifications. The lesion was measured 14×15×15 cm (Figure 1A). It was located posteriorly, involving the psoas muscles, lumbar square and subcutaneous cellular tissues on both sides, predominantly on the right. It displaced the abdominal aorta, liver and other structures of the abdominal cavity anteriorly.

CT scan showed lytic involvement of the vertebral bodies from T11-L1 and the last rib on the right side. In addition, there were multiple paraaortic lymphadenopathies of less than 1 cm in size (Figure 1B).

The patient underwent final needle aspirate (FNA) of the tumor. The results showed abundant homogeneous-looking oval cells with acidophilic and extensive cytoplasm. (Figure 2A) The cytological smears showed multiple cells with large cytoplasm, many of them multivacuolated, with a central nucleus or binucleated. This cell population showed great lack of cohesion; the cells were in a loose background of myxoid-mucoid basophilic matrix that did not seem to be in direct relation with the cells. These cells were identified as physaliferous cells (Figure 2B).

Immunohistochemistry was positive for pancreatin, EMA, Vimentin, and S-100. According to the above-mentioned findings, thoracolumbar chordoma was diagnosed for the patient. The treatment was started with Temozolomide 250 mg for 5 days, Imatinib mesylate 400 mg orally for 20 days and Erythropoietin 4000 units subcutaneously every 3 weeks. The patient did not return to the scheduled appointments, so the follow-up could not be performed. Informed consent was obtained from the patient prior to reporting the case.

Discussion

Chordoma is a very rare neoplasm. There is no incidence data for this neoplasm in Peru. Its preferred location is sacrococcygeal region, but about 15% are located in the mobile spinal column and the presentation in the thoracolumbar region is very rare.[2, 3]

The clinical manifestations of this tumor are variable. In this case, the patient presented with paraplegia due to medullary compression at the thoracolumbar level of the neoplasm. Involvement of the adjacent tissues generates pain, which is the main symptom in patients with chordoma.[7] Repeated urinary infections are frequent in patients with spinal cord injuries. This is because of alterations in the bladder ureter function, leading to reflux that predisposes patients to complicated urinary tract infections.[11]

Due to the location of the lumbar chordomas, they are easily confused with classic tumors of that region such as cystic bone aneurysms, giant cell tumors, hemangiomas, myelomas, or metastases.[1] The approximate length of the chordoma in our patient was 15×18 centimeters, found

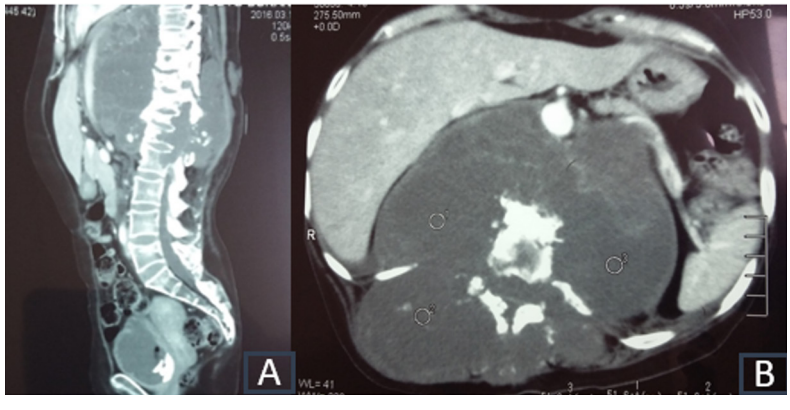

Figure 1: A) Sagittal section reveals commitments of T11, T12 and L1 B) transverse plane reveals dimensions and consistency of the expansive process

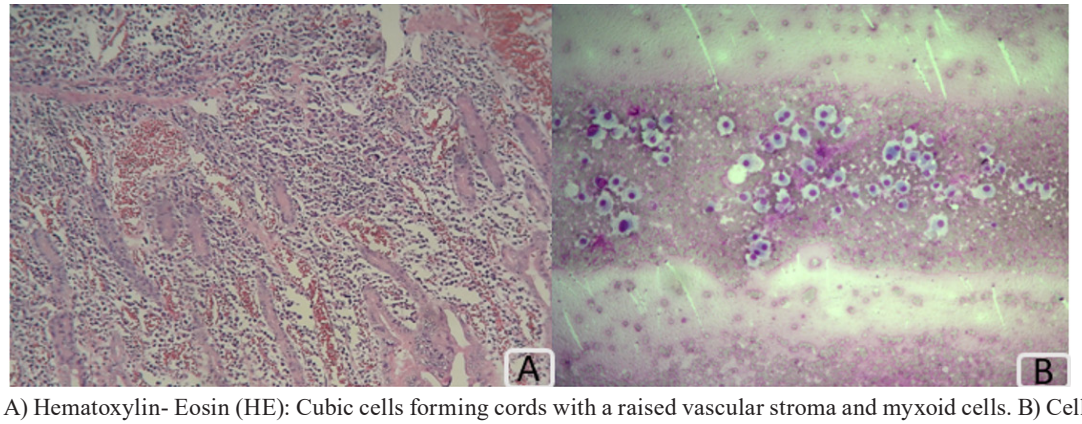

Figure 2: A) Hematoxylin- Eosin (HE): Cubic cells forming cords with a raised vascular stroma and myxoid cells. B) Cellular nests with abundant connective tissue and presence of physaliferous cells.

by the clinical inspection of the patient and according to the tomography. The tumor size was larger than the giant chordoma that is reported to be 7.8×5.6×12.1cm from China.[5] Another chordoma reported at the thoracic level had a size of approximately 2.5×2 cm.[12] Therefore, at the level of the thoracic spine, our case is considered as a giant chordoma.

Immunohistochemistry of the chordoma in our case showed the presence of the S-100 and cytokeratin. These markers are usually positive in undifferentiated chordomas with sarcomatous patterns.[13] The initial treatment of the spinal chordoma consists of local excision as soon as possible. Complete resection of the chordoma is usually difficult to perform due to vascular or neural compromise, so most of the time, the patient needs radiotherapy following the surgery. In the presented giant chordoma, surgery was not an option due to the tumor size.[14] Radiotherapy is a controversial option in these patients. A systematic review found that proton image-guided radiotherapy (IGRT) has shown good results in preoperative patients, but other modalities of radiotherapy should be discussed in order to use.[15]

Regarding systemic therapies focused on molecular markers, imatinib is one of the most common systemic therapies used in chordomas. Monotherapy is the most commonly used, where combined therapy is the second line of the treatment.[16] Other therapies such as sunitinib and erlotinib also have been suggested.[17] The combination of temozolomide and imatinib to treat chordoma is undergoing in clinical trials currently. There are phase 2 studies in patients with a 66% response and a 9-month survival.[17, 18] Our patient received this protocol, but could not be evaluated later due to loss of follow up.

A study in 138 patients showed that only 15% of patients had chordoma in lumbar spine, the mean survival time for these patients was 10 years with different treatments.[19]

Another institution has reported 12 cases of chordoma, since 1994 to 2016, in which 8 patients were alive after 5 years of follow up, and only 1 patient was alive after 13 years.[20]

Conclusion

Chordoma is a very rare neoplasm which involvement in the lumbar region is very scarce, but it should be considered in the differential diagnosis of lesions that affect the spine. Patients with earlier diagnosis would have better prognosis and could have more alternatives for treatment.

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Informed Consent

The authors obtained informed consent from the patient.

Conflict of Interest: None declared.

References

1. Bakker SH, Jacobs WCH, Pondaag W, Gelderblom H, Nout RA, Dijkstra PDS, et al. Chordoma: a systematic review of the epidemiology and clinical prognostic factors predicting progression-free and overall survival. Eur Spine J. 2018; 27(12):3043-58. doi: 10.1007/s00586-018-5764-0. PubMed PMID: 30220042.
2. Beccaria K, Sainte-Rose C, Zerah M, Puget S. Paediatric chordomas. Orphanet J Rare Dis. 2015; 10:116. doi: 10.1186/s13023-015-0340-8.
3. Youssef C, Aoun SG, Moreno JR, Bagley CA. Recent advances in understanding and managing chordomas [version 1; peer review: 2 approved]. F1000Research 2016, 5(F1000 Faculty Rev):2902. doi: 10.12688/f1000research.9499.1
4. Barrenechea IJ, Perin NI, Triana A, Lesser J, Costantino P, Sen C. Surgical management of chordomas of the cervical spine. J Neurosurg Spine. 2007; 6(5):398-406. doi: 10.3171/spi.2007.6.5.398. PubMed PMID: 17542504.
5. Pu F, Wang B, Liu J, Chen F, Shao Z. Giant chordoma in the thoracolumbar spine: a case report and literature review. Eur Spine J. 2017; 26(Suppl 1):95–9. doi: 10.1007/s00586-016-4877-6. PubMed PMID: 27900552.
6. Elefante A, Caranci F, Del Basso De Caro ML, Peca C, Guadagno E, Severino R, et al. Paravertebral high cervical chordoma. A case report. Neuroradiol J. 2013; 26(2):227–32. doi: 10.1177/197140091302600214. PubMed PMID: 23859247.
7. Williams BJ, Raper DM, Godbout E, Bourne TD, Prevedello DM, Kassam AB, et al. Diagnosis and treatment of chordoma. J Natl Compr Canc Netw. 2013; 11(6):726–31. doi: 10.6004/jnccn.2013.0089. PubMed PMID: 23744869.
8. Alshammari J, Monnier P, Daniel RT, Sandu K. Clival chordoma with an atypical presentation: a case report. J Med Case Rep. 2012; 6(1):410. doi:10.1186/1752-1947-6-410
9. Boari N, Gagliardi F, Cavalli A, Gemma M, Ferrari M, Riva P, et al. Skull base chordomas: clinical outcome in a consecutive series of 45 patients with long-term follow-up and evaluation of clinical and biological prognostic factors. J Neurosurg. 2016;125(2):450–60. doi: 10.3171/2015.6.JNS142370.
10. Mima M, Demizu Y, Jin D, Hashimoto N, Takagi M, Terashima K, et al. Particle therapy using carbon ions or protons as a definitive therapy for patients with primary sacral chordoma. Br J Radiol. 2014; 87(1033):20130512.
11. Brommer B, Engel O, Kopp MA, Watzlawick R, Müller S, Prüss H, et al. Spinal cord injury-induced immune deficiency syndrome enhances infection susceptibility dependent on lesion level. Brain. 2016; 139(Pt 3):692–707.
12. Vaidya MM, Shenoy AS, Goel NA. Chondrosarcoma of the dorsal spine - A rare case. J Craniovertebr Junction Spine. 2019; 10(4):250–3. doi: 10.4103/jcvjs.JCVJS_100_19. PubMed PMID: 32089620. PubMed

Central PMCID: PMC7008663.
13. Ridenour RV 3rd, Ahrens WA, Folpe AL, Miller DV. Clinical and histopathologic features of chordomas in children and young adults. Pediatr Dev Pathol. 2010; 13(1):9-17. doi: 10.2350/09-01-0584.1. PubMed PMID: 19348512.
14. Stacchiotti S, Casali PG, Lo Vullo S, Mariani L, Palassini E, Mercuri M, et al. Chordoma of the mobile spine and sacrum: a retrospective analysis of a series of patients surgically treated at two referral centers. Ann Surg Oncol. 2010; 17(1):211-9. doi: 10.1245/s10434-009-0740-x.
15. Pennicooke B, Laufer I, Sahgal A, Varga PP, Gokaslan ZL, Bilsky MH, et al. Safety and Local Control of Radiation Therapy for Chordoma of the Spine and Sacrum: A Systematic Review. Spine (Phila Pa 1976). 2016; 41 Suppl 20(Suppl 20):S186-S192.
16. Meng T, Jin J, Jiang C, Huang R, Yin H, Song D, et al. Molecular Targeted Therapy in the Treatment of Chordoma: A Systematic Review. Front Oncol. 2019; 9:30. doi: 10.3389/fonc.2019.00030. PubMed PMID: 30775316.
17. Colia V, Provenzano S, Hindi N, Casali PG, Stacchiotti S. Systemic therapy for selected skull base sarcomas: Chondrosarcoma, chordoma, giant cell tumour and solitary fibrous tumour/hemangiopericytoma. Rep Pract Oncol Radiother. 2016; 21(4):361-9. doi: 10.1016/j.rpor.2015.12.005. PubMed PMID: 27330421.
18. Schuetze SM, Bolejack V, Choy E, Ganjoo KN, Staddon AP, Chow WA, et al. Phase 2 study of dasatinib in patients with alveolar soft part sarcoma, chondrosarcoma, chordoma, epithelioid sarcoma, or solitary fibrous tumor. Cancer. 2017; 123(1):90–7. doi: 10.1002/cncr.30379. PubMed PMID: 27696380.
19. Hindi N, Casali PG, Morosi C, Messina A, Palassini E, Pilotti S, et al. Imatinib in advanced chordoma: A retrospective case series analysis. Eur J Cancer. 2015; 51(17):2609–14. doi: 10.1016/j.ejca.2015.07.038. PubMed PMID: 26283036.
20. D'Amore T, Boyce B, Mesfin A. Chordoma of the mobile spine and sacrum: clinical management and prognosis. J Spine Surg. 2018; 4(3):546-52. doi: 10.21037/jss.2018.07.09. PubMed PMID: 30547117. PubMed Central PMCID: PMC6261773.