GSC Advanced Research and Reviews
eISSN: 2582-4597 CODEN (USA): GARRC2
Cross Ref DOI: 10.30574/gscarr
Journal homepage: https://gsconlinepress.com/journals/gscarr/

(CASE REPORT)

Sacral chordoma with pulmonary metastasis: Case report and a brief review of the literature

Alexa Kessen [2], Oliver Stewart [3], Marina Moussa [2], Ross Jenkins [2], Pedro Novo [4], Zainab Jasim [5], Arline Ficarra [2], Jessica Jahoda [1,2] and Mohamed Aziz [1,*]

[1] *Research Writing & Publication (RWP), LLC, NY, USA.*
[2] *American University of the Caribbean School of Medicine, USA.*
[3] *Ross University School of Medicine, Barbados.*
[4] *St. George's University School of Medicine, Grenada.*
[5] *UMHS University of Medicine and Health Sciences, St. Kitts.*

GSC Advanced Research and Reviews, 2025, 24(03), 054–062

Publication history: Received on 25 July 2025; revised on 29 August 2025; accepted on 02 September 2025

Article DOI: https://doi.org/10.30574/gscarr.2025.24.3.0265

Abstract

Chordoma is an uncommon, locally invasive malignancy that originates from residual notochords, and the sacral location constitutes half of the cases. Despite the multimodal treatment, the treatment becomes convoluted due to anatomical limitations and recurrence rates. We report a 48-year-old man with an 8-month history of progressive sacrococcygeal pain and neurological manifestations—initial presentation involved nocturnal exacerbation of pain, bowel/bladder dysfunction, and weakness of lower extremities. On physical examination, decreased sacral reflexes and palpation of a presacral mass on digital rectal examination were noted. MRI revealed a lobulated, T2-hyperintense mass in the sacrum, with extension into the surrounding soft tissues. Chordoma was confirmed by CT-guided core biopsy, which revealed typical physaliferous cells, positive immunostaining for brachyury, and expression of PDGFR.

After multidisciplinary tumor board discussion, the patient was treated with surgical resection of a 6 cm tumor with focally positive margins, followed by concurrent high-dose radiotherapy (76 Gy) and imatinib mesylate (400 mg/d). Even though the patient responded initially, he developed pulmonary metastasis 19 months later. He refused surgical treatment or radiation therapy and accepted only medical treatment, to which he responded with serious side effects, and expired four months later due to widespread metastasis. The case illustrates the multifaceted approach to managing sacral chordoma, highlighting the importance of early diagnosis, multidisciplinary care coordination, and aggressive multimodal treatment. Molecular profiling and targeted therapy integration are treatment modalities that are still in the process of development. Although management is at its best, the emergence of treatment resistance underscores the need for innovative treatment approaches and the vital role of long-term surveillance in chordoma patients.

Keywords: Chondroma; Sacrum; Malignant; Metastasis; Molecular

1. Introduction

Chordoma is an uncommon notochordal malignant bone tumor present in 1-4% of primary bone malignancies. Although its incidence is very low at 0.08/100,000 population per year, it is the most prevalent primary tumor of the sacrum. [1] Nearly half of the cases are affected in the sacrococcygeal area, and then the skull base and mobile spine. The tumor tends to be indolent but more locally invasive and is very challenging to diagnose and treat. [2]

* Corresponding author: Mohamed Aziz.

Clinically, sacral chordomas present late due to the deep location and large potential space in the presacral region. Patients frequently complain of chronic lumbosacral back pain, radiculopathy, bowel, and bladder effects that resemble degenerative or inflammatory spinal diseases, and cause a delay in diagnosis. [3] Imaging finding of a destructive midline mass with lobulated margins are typical and confirmed by histopathology, where the physaliferous cells and expression of brachyury are seen. Brachyury is an embryonic transcription factor that promotes cell migration and is crucial for the development of the notochord and posterior mesoderm. [4][5]

The task of management is multidisciplinary. Extensive surgical resection with negative margins is the mainstay of treatment, but is often limited by the proximity to sacral nerve roots and other pelvic structures. External radiotherapy and targeted therapies, especially those that inhibit platelet-derived growth factor receptor (PDGFR) and its signaling pathways (tyrosine kinase inhibitors), are increasingly used to enhance local disease management and delay disease progression. [5]

This case introduces a 48-year-old male patient with sacral chordoma who has acquired pulmonary metastases despite the use of multiple modalities, and we briefly review the literature to demonstrate the current knowledge on its pathogenesis, diagnosis, and management modalities.

2. Case presentation

A 48-year-old man presented to the clinic with an 8-month history of progressive lower back pain. Pain was initially random and described as occupational strain, but over time, it became more persistent and intense, characterized by a deep, aching sensation localized to the sacrococcygeal area. The pain followed a characteristic pattern, worsening at night and when lying down, and it did not improve with rest, unlike mechanical back pain. The patient experienced gradual neurological symptoms, including numbness of both lower limbs, plantar flexion weakness, and bowel dysfunction, which was characterized by constipation and long urination. Conservative management with NSAIDs and physical therapy provided only temporary relief, leading to a delay in diagnosis. On physical examination, the Achilles reflexes were reduced on both sides, there was loss of sensation in S1-S5 dermatomes, and plantar flexion was weak. A firm and fixed presacral mass was detected by digital rectal examination. The patient reported no personal or family history of malignancy or other significant medical conditions. Laboratory findings including CBC, ESR, and CRP were normal. At this point, the differential diagnosis was mechanical lumbar pathology, infectious disease (osteomyelitis), and neoplastic disease (primary bone tumors, metastatic disease). Based on the clinical data, an MRI of the lumbosacral spine was recommended to characterize the lesion further and evaluate neural involvement.

On plain radiographs of the lumbosacral spine, there was mild osteolytic destruction of sacral segments, absence of normal sacral curvature, and the possibility of a soft tissue extension. The CT scan revealed destruction of the sacral bones accompanied by calcifications. A mass about 6 cm in size was detected on MRI studies. The tumor was hypointense to isointense compared to muscle on T1-weighted images. On T2-weighted sequences, the appearance was lobulated and honey-combed with heterogeneous high signal intensity indicative of myxoid matrix and cystic elements. The pattern of enhancement was heterogeneous, exhibiting both peripheral and septal enhancement patterns. The tumor extended both anteriorly into the presacral space and posteriorly, with large components of soft tissues. The radiological differential involved several entities that shared the same features. Giant cell tumors can exhibit similar osteolytic changes, but in most cases, it is more homogeneous in enhancement and do not typically show T2 hyperintensity. Metastases, especially those of renal cell carcinoma, may resemble the imaging appearance of chordoma, but tend to be multiple. The most similar differential is chondrosarcoma, which exhibits similar T2 features but typically has more conspicuous calcifications and anatomical predilection sites.

Percutaneous core needle biopsy by the posterior approach guided by CT was performed. Histologically, the tumor consisted of lobules of cells separated by fibrous septa, exhibiting a lobulated growth pattern. The dominant cellular element was a large, vacuolated cell with abundant eosinophilic to clear cytoplasm and an eccentric nucleus. An abundant extracellular myxoid matrix surrounded these cells. The cell population was dimorphic, consisting of small epithelioid cells and larger vacuolated, foamy cells. Nuclear pleomorphism was mostly mild to moderate, and mitotic activity was low. There were occasional foci that were highly pleomorphic. (Figure 1 A, B, C, D) The major differential considerations were chondrosarcoma, especially the myxoid types, which can exhibit a similar myxoid matrix but do not show large vacuolated foamy cells and display different immunohistochemical characteristics. Sometimes, metastatic adenocarcinoma (usually of genitourinary or gastrointestinal origin) can display similar features; however, these are typically multiple and should exhibit a distinct immunoprofile.

The diagnosis was established through immunohistochemistry (IHC) studies. There was positive brachyury, a T-box transcription factor (a group of transcription factors involved in embryonic limb and heart development). Additionally,

IHC markers were positive for cytokeratins (CK8, CK18, CK19), EMA, and showed focal positivity for S-100 protein. The tumor cells were negative for SOX-9, CDX-2, and PSA. The presence of brachyury, in conjunction with the expression of cytokeratin in a morphologically correct context, confirmed the diagnosis of sacral conventional chordoma. Molecular profiling can reveal multiple targetable pathways for therapeutic intervention, so molecular studies were conducted. Tumor molecular analysis revealed that the tumor expresses PDGFR, which is why imatinib and other similar agents may be therapeutic options. Body and bone scans, and no evidence of metastasis, confirmed a localized tumor in the sacrum with extension to the soft tissue.

Sacral chordoma was a complex diagnosis that required multidisciplinary assessment by orthopedic oncology, neurosurgery, radiation oncology, medical oncology, and pathology expert review. The tumor resectability, its proximity to vital neural structures, the patient's age and functional status, and the trade-offs between oncological and functional preservation principles were the major considerations in the discussion. The expression of PDGFR added some therapeutic considerations that further affected the general treatment plan. A wide margin en bloc surgical resection was performed, aiming to obtain wide negative margins whenever anatomically possible; a combined anterior-posterior surgical approach was employed. Nevertheless, the anatomical limitations of the sacrum, especially the vicinity to vital neural structures and the sacral plexus, required a compromise between oncological sufficiency and the preservation of function. The 6 x 5 cm tumor mass was removed, but the tumor involved foci of the surgical margins.

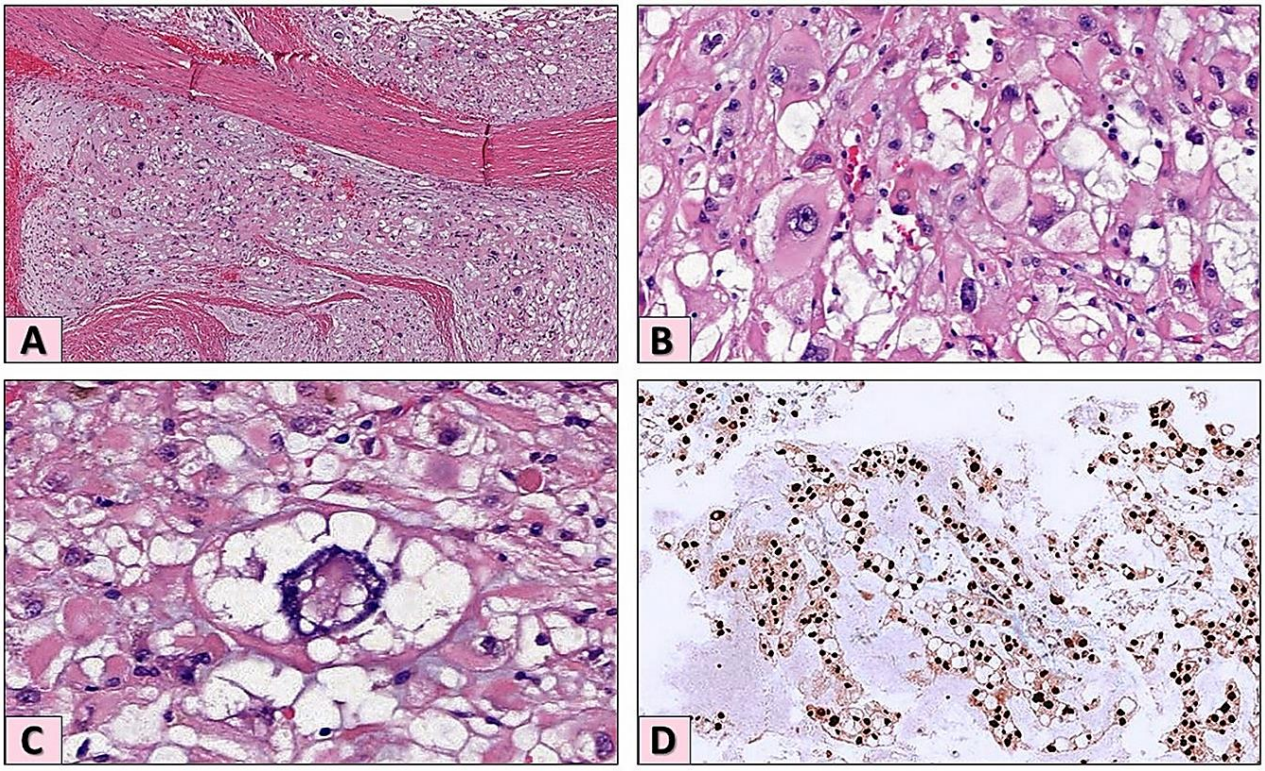

Figure 1 Histomorphology of sacral chordoma

1A Low power view showing lobules of cells separated by fibrous septa, showing a lobulated growth pattern (H&E stain X20); **1B:** High power view showing dimorphic cell population, with small epithelioid cells and the larger vacuolated foamy cells. Nuclear pleomorphism is mostly mild to moderate, and mitotic activity was low (H&E stain X40); **1C:** High power view showing the foamy, vacuolated, characteristic physaliferous cell (H&E stain X60); **1D**: Tumor cells positive for S100

Postoperative high-dose radiation therapy through intensity-modulated radiation therapy (IMRT) was employed. The radiation dose was between 70 and 80 Gy, which was administered in conventional fractionation. Imatinib therapy, 400-800 mg daily, was also added concurrently. The patient underwent close monitoring, with functional deficits addressed as they arose. Imaging follow-up was performed every 3-4 months because of the high risk of local recurrence. A chest CT scan at 19 months after the first treatment revealed three pulmonary nodules: a left-sided lung nodule (2.1 cm and 1.8 cm) and a right-sided lung nodule (3.0 cm). CT-guided biopsy of the largest lesion identified metastatic chordoma with the same histological characteristics as the primary tumor, positivity of the brachyury, and preservation of PDGFR expression. Re-staging assessment showed no signs of further metastatic sites, but the primary site disease was stable. The patient opted for a non-surgical approach and declined further radiation therapy after all therapeutic

options were discussed. He received systemic therapy change to sunitinib 50 mg daily (4 weeks on/ 2 weeks off) with clinical trial enrollment in mind. The patient had a severe side effect response to the medical intervention and died four months after the metastasis was diagnosed, due to widespread metastasis to the lung, bone, skin, and soft tissue.

3. Discussion

3.1. History and WHO classification

Chordomas are rare malignant tumors that develop from the remnants of the embryonic notochord and were first described by Virchow in 1857, followed by further characterization by Ribbert in 1894. [6] They comprise about 1-4% of the total primary malignant bone tumors, and occur at 0.08 per 100,000 population annually. [7] The most common site of involvement is the sacrum, where almost half of all chordomas occur, followed by the skull base (clivus) and mobile spine. Their slow, yet locally aggressive growth pattern, in addition to a high rate of recurrence, has long rendered them a therapeutic challenge. [1]

Clear environmental or lifestyle risk factors for chordoma have not been identified. Most cases are sporadic, with some family clustering described as rare. Genetic research has discovered that germline duplications of the T gene (brachyury) on chromosome 6q27 are a risk factor in familial cases of the condition. [6] Chordomas are common in adults aged between the fourth and seventh decades of life, with a slight male predisposition. Pediatric cases are uncommon and often associated with atypical histological variants. [7] The clinical manifestation is determined by the tumor location and local invasion, and the lesions of the sacral area tend to cause pain, neurologic deficits, bowel, or bladder dysfunction, as in our patient. [3]

The 2020 list of bone and soft tissue tumors by the World Health Organization (WHO) defines chordomas as malignant tumors of notochordal origin, and further classifies them into three major histological types: conventional (classic), chondroid, and dedifferentiated chordoma. The most common is conventional chordoma, as in our case, and is typified by physaliferous cells in a myxoid stroma. Chondroid chordoma presents regions that resemble hyaline cartilage and may be associated with a slightly better prognosis. The least common and most aggressive, dedifferentiated chordoma, has biphasic morphology with high-grade sarcomatous transformation and a dismal prognosis. [8] [9] This pathological, genetic, and historical framework provides an emphasis on the complexity of the biology of chordomas, emphasizing the need to classify these tumors correctly to support prognosis and treatment options.

3.2. Clinical presentation and Imaging findings

A sacral chordoma with pulmonary metastasis typically presents with lower back pain, radiculopathy, neurological deficits, bladder and bowel dysfunction, and metastatic symptoms. [9] Lower back pain is the most common initial symptom, often presenting around the decade of life. [10], as our patient presented with lower back pain as his initial complaint. The pain can be described as dull or achy and worsens with prolonged sitting or lying. [11] Radiculopathy presents with pain radiating down the leg due to nerve compression. [12] This condition can present as unilateral or bilateral and may radiate to the buttocks, posterior thigh, or leg, ultimately affecting gait. [11] Compression of nerve roots can lead to weakness or loss of function in the lower extremities. Sacral chordoma can impact bowel and bladder function, leading to constipation, urinary tract infections, urinary retention, or incontinence. [10] [11] Many patients may initially be misdiagnosed or treated for other conditions (back pain, sciatica) before a correct diagnosis is made, as is the case with our patient. CT and MRI are used to determine the extent of chordoma lesions and aid in surgical planning; however, pathological examination following tissue biopsy remains the gold standard for diagnosing sacral chordoma. (12]

Metastasis is relatively uncommon, with the lung representing the most common site. [13] A 2024 study from Perking University analyzed patients with sacral chordoma and found that tumor size, recurrence after surgery, and prior sacral operations were associated with an increased risk of lung metastases. The overall survival in patients with lung metastases was greatly lower than in those with no metastases. Although radiotherapy was linked to the occurrence of lung metastasis, radiotherapy was also found to be a protective factor against lung metastasis in certain settings and a sample study showed lung metastasis to be a negative prognostic factor with respect to overall survival among patients with sacral chordoma. [3] The diagnosis of lung metastasis is primarily established through chest CT scans, although few patients undergo surgical excision for pathological confirmation. [14]

3.3. Diagnosis: Pathological and molecular considerations

Chordoma diagnosis involves considering clinical suspicion and imaging, with a confirmatory histopathological diagnosis. Regular lab tests are generally unimpressive; inflammatory biomarkers are generally normal, and serum

tumor markers are not present to substantiate the diagnosis. Therefore, laboratory testing plays a minimal role, except in the diagnosis and treatment of infections or systemic malignancies. [1]

The definitive diagnosis rests on tissue sampling. Image-guided core needle biopsy is the method of choice because it yields sufficient tissue for histological study and additional tests, while minimizing contamination of the adjacent planes. The sensitivity of fine-needle aspiration (FNA) is low due to the gelatinous character of the tumor and the poor maintenance of architectural patterns. [15] In our instance, a CT-guided core biopsy had furnished diagnostic material and had prevented avoidable surgical morbidity.

The classical appearance of histopathology includes lobules of tumor cells divided by fibrous septa. Characteristics include large cells with vacuolated cytoplasm, eccentric nuclei, and physaliferous cells embedded in a myxoid matrix. Tumor cellularity is unpredictable, and although mitotic figures are usually sparse, focal pleomorphism can occur. Chondroid chordomas exhibit cartilaginous-like areas, and their dedifferentiated variants display a sudden, progressive transformation into high-grade sarcoma. [4] [7]

The differential diagnosis is wide-ranging with the presence of chondrosarcoma (especially the myxoid type), metastatic adenocarcinoma (usually of gastrointestinal or genitourinary origin), and, albeit infrequently, other myxoid tumors of soft tissue. Radiologic correlation and immunohistochemistry (IHC) play a crucial role in reducing diagnostic discrepancies. [15]

Immunohistochemistry is central. Epithelial and mesenchymal markers, such as cytokeratins (CK8/18/19), EMA, and S-100 protein, are normally co-expressed by chordomas. [2] The most valuable test is brachyury, which is a nuclear transcription factor coded by the T gene, and is positive in almost all traditional and chondroid chordomas. This contrasts with chordoma, chondrosarcoma (SOX-9 positive, brachyury negative), and metastatic adenocarcinomas (positive for lineage-specific markers, e.g., CDX2 or PSA, and negative for brachyury). [15]

Molecular studies further refine molecular diagnosis and treatment advice. Changes in the PI3K/AKT/mTOR pathway and the expression of PDGFR have therapeutic implications, justifying the use of targeted agents such as imatinib or tyrosine kinase inhibitors. The rare poorly differentiated subtype is characterized by loss of SMARCB1/INI1 and is commonly seen in younger patients. [17] [18]

After histopathological and molecular validation, these cases should be discussed in a multidisciplinary tumor board, where surgical, radiation, and systemic therapies are evaluated in relation to anatomical limitations and molecular targets. In our patient, the diagnosis of classical sacral chordoma with PDGFR expression directly influenced the treatment strategy, leading to the addition of postoperative imatinib.

3.4. Treatment Strategies

The management of chordoma remains one of the most challenging areas in musculoskeletal oncology due to the tumor's deep location, proximity to vital structures, and high recurrence rates. [5] The cornerstone of treatment is surgical resection with wide negative margins, which offers the best chance for local control and prolonged survival. However, achieving clear margins in the sacral region is particularly difficult, as en bloc resection often necessitates the sacrifice of sacral nerve roots, leading to significant morbidity with bowel, bladder, and sexual dysfunction. [16] In our patient, a wide-margin resection was attempted, but anatomical constraints resulted in focally positive margins, a common limitation in sacral chordoma surgery.

Radiotherapy plays a critical role as an adjunct or alternative when negative margins cannot be achieved. Conventional external-beam radiation has limited efficacy due to chordoma's relative radioresistance. However, high-dose conformal techniques such as intensity-modulated radiation therapy (IMRT), proton beam therapy, and carbon ion therapy have significantly improved local control. [17]

Systemic therapy has historically had little role, as chordomas are generally resistant to standard cytotoxic chemotherapy. Increasingly, attention has shifted toward molecularly targeted therapies. [17] [18] The expression of platelet-derived growth factor receptor (PDGFR), epidermal growth factor receptor (EGFR), and activation of the PI3K/AKT/mTOR pathway have provided rational targets. Tyrosine kinase inhibitors (TKIs) such as imatinib, sunitinib, and sorafenib have shown variable responses in advanced or metastatic disease, primarily offering disease stabilization rather than a cure. [17] [18] In our case, imatinib was incorporated into adjuvant therapy due to PDGFR expression, with subsequent transition to sunitinib upon pulmonary metastasis—unfortunately, systemic toxicity limited further benefit.

Emerging approaches include immune checkpoint inhibitors, brachyury-directed vaccines, and combination regimens, which are under investigation in clinical trials. Although these strategies remain experimental, they highlight the ongoing shift toward personalized medicine in chordoma management. [19] Ultimately, optimal outcomes depend on multimodal therapy and long-term surveillance. Even with aggressive treatment, local recurrence and distant metastasis remain significant risks, as illustrated by our patient's pulmonary metastases. This highlights the importance of coordinated care within specialized centers, the early integration of molecular profiling, and ongoing research into novel therapeutics.

3.5. Prognosis and Outcomes

A paradoxical clinical behavior characterizes chordomas: they grow slowly but relentlessly, with a marked tendency for local recurrence and eventual metastasis to distant sites. [3] The natural history is shaped largely by the adequacy of local control, as complete surgical excision with negative margins remains the most important determinant of outcome. [5] Even with aggressive management, however, recurrence rates remain high, ranging from 30–60% within five years after treatment. Positive surgical margins, as in our patient, significantly increase the risk of early recurrence. [7]

Metastasis occurs in approximately 20–40% of patients, most commonly involving the lungs, followed by bone, liver, and lymph nodes. Pulmonary metastasis, as seen in our case, is the typical first site and may remain indolent for a considerable period. [1] However, once systemic dissemination is established, survival outcomes decline sharply, as effective systemic therapies are limited. [6]

Overall survival in chordoma has improved over recent decades, thanks to advances in surgical techniques and radiotherapy. Reported five-year survival rates range from 50–70%, while ten-year survival rates decline to 35–50%, [3] reflecting the cumulative impact of local recurrence and metastasis. Prognosis varies by subtype: conventional and chondroid chordomas tend to follow a more protracted course, whereas dedifferentiated and poorly differentiated variants exhibit aggressive behavior with median survival measured in months. [4]

Several prognostic factors influence outcomes. Favorable predictors include younger age, smaller tumor size, sacral location below S3, and successful wide-margin resection. Conversely, high tumor grade, dedifferentiated histology, and incomplete resection portend a worse prognosis. [2] Molecular features are also emerging as prognostic indicators: brachyury expression confirms diagnosis but does not independently predict outcome, while loss of SMARCB1/INI1 in poorly differentiated chordoma is associated with aggressive disease. [20]

In clinical practice, prognosis encompasses not only biological factors but also functional aspects of the disease. Sacral resections may achieve disease control at the cost of significant morbidity, and patient quality of life must be balanced against oncological goals. [5] In our patient, despite aggressive multimodality therapy, pulmonary metastasis developed within two years, and systemic therapy was limited by toxicity, reflecting the sobering reality of current therapeutic limitations.

Ultimately, chordoma remains a chronic, relapsing malignancy, where the emphasis lies in achieving maximal safe resection, consolidating with high-dose radiotherapy, and maintaining vigilant long-term surveillance. Future improvements in survival will likely depend on the integration of molecularly targeted and immunotherapeutic strategies with conventional modalities.

3.6. Pathogenesis and pathophysiology

Chordomas develop out of remnants of embryonic notochordal cells, which remain in the axial skeleton. Such cells are normally regressed during development; however, in very rare cases, they survive and can undergo malignant transformation. [18] The key aspect of this is the transcription factor, brachyury, coded by the T gene on chromosome 6q27, which is required for the development of the notochord. Only a hereditary predisposition to chordoma has been identified, specifically the duplication of this gene, and overexpression of brachyury is a characteristic finding in virtually all cases. [21]

Many cases of chordomas exhibit recurrent loss of heterozygosity on chromosome 3p, which in turn leads to downstream effects of the SETD2 gene. This gene encodes a histone methyltransferase responsible for modifying H3K36me3. This dysregulation may correlate with clinical outcomes. [21] [22]

On a cellular level, chordomas are characterized by slow proliferation and invasive expansion, indicating the activation of pathways that enhance survival over high mitotic activity. The PDGFR, EGFR, and PI3K/AKT/mTOR signaling pathways aberrantly promote tumor cell survival, angiogenesis, and anti-apoptotic protection. Their gelatinous and

lobulated shape, as well as the proliferation by infiltration of tissue space, are aided by the richness of their myxoid extracellular matrix, thus posing a real challenge to resection. [23]

These biological characteristics clarify the clinical behavior of chordomas, including slow progression, common local recurrence, and late metastasis, as well as the disease's therapeutic resistance to traditional chemotherapy and low-dose radiotherapy. [18] innovations in molecular profiling are, therefore, not only enhancing the accuracy of diagnosis but also improving the development of targeted and immunotherapeutic approaches to circumvent natural mechanisms of resistance.

3.7. What have we learned from this case?

This case highlights several key points regarding the management of sacral chordoma. To begin with, the clinical manifestations are frequently insidious and nonspecific, leading to a late diagnosis. The patient's mechanical causes of persistent back pain and neurological dysfunction were the initial factors identified in our patient. As such, it is important to suspect mechanical causes early in the course of recent symptoms, particularly pain that is progressive, nocturnal, or accompanied by neurological deficits.

Second, despite the evolution of multimodal therapy, such as surgery, high-dose radiation, and targeted therapy, local recurrence and metastatic progression are frequent issues. Achieving favorable surgical margins in the sacral region is often unattainable, which further increases the risk of recurrence.

Third, molecular profiling is becoming more imperative, not only to have diagnostic accuracy, but also to inform systemic therapy. Here, the expression of PDGFR made it possible to use imatinib and subsequently sunitinib, but treatment was eventually restricted due to its toxicity and the progression of disease.

Ultimately, this case has demonstrated that multidisciplinary tumor boards play a significant role in balancing oncological management and functional survival, and that long-term follow-up is essential. Pulmonary metastases development demonstrates that even with aggressive treatment, the current practices are still constrained and that new treatment methods are urgently required.

4. Conclusion

Sacral chordoma is an uncommon malignancy that poses significant challenges, including diagnostic delays, complex surgical anatomy, and limited systemic therapies. Reporting on this case contributes to the growing body of evidence on the importance of early recognition, multidisciplinary collaboration, and the incorporation of molecular diagnostics. In addition to the clinical specifics, this case raises the broader issue of improving outcomes through innovative treatment strategies and careful long-term follow-ups.

Compliance with ethical standards

Acknowledgments

Special thanks to Michael Mitchell, Natalia Berio-Perez, and Kassandra Piris for their assistance in reviewing the final manuscript.

Disclosure of conflict of interest

All authors make the following declarations:

- **Payment/services information:** All authors have declared that they received no financial support from any organization for the submitted work.
- **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might be interested in the submitted work.

Statement of ethical approval

Ethical review and approval were not required for this study involving human participants. The paper has been sufficiently anonymized to maintain the patient's confidentiality.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

Statement of informed consent

The patient expired, and all attempts to reach the family members were unsuccessful. Therefore, the paper has been sufficiently anonymized to maintain patient confidentiality.

References

[1] Cheryl Ann Palmer MD, Cheryl Ann Palmer MD, Eric A Goold MD. Chordoma. Medscape. Updated: May 03, 2022, accessed August 28, 225. Available at https://emedicine.medscape.com/article/250902-overview?utm_source=chatgpt.com

[2] Karele EN, Paze AN. Chordoma: To know means to recognize. Biochimica et Biophysica Acta (BBA)-Reviews on Cancer. 2022 Sep 1;1877(5):188796.

[3] Shi Q, Guo W, Yu S, Xu J, Ji T, Tang X. Clinical characteristics and predisposing factors of lung metastasis in sacral chordoma: a cross-sectional cohort study of 221 cases. Frontiers in Oncology. 2024 Aug 12;14:1416331.

[4] Desai R, Pelargos PE, Dunn IF. Chordoma: genetics and contemporary management. International Journal of Molecular Sciences. 2024 May 28;25(11):5877.

[5] Barber SM, Sadrameli SS, Lee JJ, Fridley JS, Teh BS, Oyelese AA, Telfeian AE, Gokaslan ZL. Chordoma—current understanding and modern treatment paradigms. Journal of clinical medicine. 2021 Mar 4;10(5):1054.

[6] Sahyouni R, Goshtasbi K, Mahmoodi A, Chen JW. A historical recount of chordoma. Journal of Neurosurgery: Spine. 2018 Apr 1;28(4):422-8.

[7] Steven Tenny, Matthew A. Varacallo. StatPearls [Internet]. Last Update: August 7, 2023. Available at https://www.ncbi.nlm.nih.g /t=Epidogy

[8] Anderson WJ, Doyle LA. Updates from the 2020 World Health Organization classification of soft tissue and bone tumours. Histopathology. 2021 Apr;78(5):644-57.

[9] Choi JH, Ro JY. The 2020 WHO classification of tumors of bone: an updated review. Advances in anatomic pathology. 2021 May 1;28(3):119-38.

[10] Pillai S, Govender S. Sacral chordoma: a review of literature. Journal of orthopaedics. 2018 Jun 1;15(2):679-84.

[11] Quraishi NA, Giannoulis KE, Edwards KL, Boszczyk BM. Management of metastatic sacral tumours. European Spine Journal. 2012 Oct;21(10):1984-93.

[12] Bai R, Zhao ZQ, Wang YX, Zhao W, Wu LS, Cui SX, Guo SB, Liang CL. Sacral and thoracic chordoma with pulmonary metastases: a case report and review of the literature. Molecular and Clinical Oncology. 2021 Jan;14(1):17.

[13] Young VA, Curtis KM, Temple HT, Eismont FJ, DeLaney TF, Hornicek FJ. Characteristics and patterns of metastatic disease from chordoma. Sarcoma. 2015;2015(1):517657.

[14] Kano H, Niranjan A, Lunsford LD. Radiosurgery for Chordoma and Chondrosarcoma. Progress in Neurological Surgery. 2019 May 16;34:207-14.

[15] Veronica Ulici MD, Hart J. Chordoma A Review and Differential Diagnosis. Archives of Pathology & Laboratory Medicine. 2022 Mar 1;146(3):386-95.

[16] Denaro L, Berton A, Ciuffreda M, Loppini M, Candela V, Brandi ML, Longo UG. Surgical management of chordoma: a systematic review. The journal of spinal cord medicine. 2020 Nov 1;43(6):797-812.

[17] Meng T, Jin J, Jiang C, Huang R, Yin H, Song D, Cheng L. Molecular targeted therapy in the treatment of chordoma: a systematic review. Frontiers in oncology. 2019 Feb 1;9:30.

[18] Jin MC, Connolly ID, Ravi K, Tobert DG, MacDonald SM, Shin JH. Unraveling molecular advancements in chordoma tumorigenesis and treatment response: a review of scientific discoveries and clinical implications. Neurosurgical Focus. 2024 May 1;56(5):E18.

[19] Walker RL, Hornicek FJ, Duan Z. Advances in the development of chordoma models for drug discovery and precision medicine. Biochimica et Biophysica Acta (BBA)-Reviews on Cancer. 2022 Nov 1;1877(6):188812.

[20] Mobley BC, McKenney JK, Bangs CD, Callahan K, Yeom KW, Schneppenheim R, Hayden MG, Cherry AM, Gokden M, Edwards MS, Fisher PG. Loss of SMARCB1/INI1 expression in poorly differentiated chordomas. Acta neuropathologica. 2010 Dec;120(6):745-53.

[21] Chen M, Wu Y, Zhang H, Li S, Zhou J, Shen J. The roles of embryonic transcription factor BRACHYURY in tumorigenesis and progression. Frontiers in Oncology. 2020 Jun 30;10:961.

[22] Zhu GG, Ramirez D, Chen W, Lu C, Wang L, Frosina D, Jungbluth A, Ntiamoah P, Nafa K, Boland PJ, Hameed MR. Chromosome 3p loss of heterozygosity and reduced expression of H3K36me3 correlate with longer relapse-free survival in sacral conventional chordoma. Human pathology. 2020 Oct 1;104:73-83.

[23] Dewaele B, Maggiani F, Floris G, Ampe M, Vanspauwen V, Wozniak A, Debiec-Rychter M, Sciot R. Frequent activation of EGFR in advanced chordomas. Clinical sarcoma research. 2011 Jul 25;1(1):4.