

Case Report

Pain in Ewing Sarcoma: A Complex Case Report

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Abstract

Ewing sarcoma is a rare bone and tissue cancer which most commonly occurs in young adults and children. Survival rates are variable, recurrences common, and premature mortality a considerable burden despite aggressive multimodal cancer therapy. Pain is frequently experienced from the time of Ewing sarcoma diagnosis and even in favourable outcomes, chronic pain syndromes are not uncommon developments. However, the literature around the pain experience and pain-related distress, along with potential management strategies, is lacking. In this case report, we describe a patient with Ewing sarcoma of the right pelvis which was later complicated by developing secondary chondrosarcoma. This report will highlight the pain characteristics, syndromes, and challenges of pain management of a patient with Ewing sarcoma spanning multiple specialties and exploring key evolving concepts in cancer pain management.

Keywords: cancer pain, Ewing sarcoma, cancer pain syndromes

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1 INTRODUCTION

Ewing sarcoma is a rare bone and tissue cancer commonly affecting children and young adults^[1]. It is the second most frequent primary bone cancer after osteosarcoma and is characterized by small round blue cell tumors on morphology^[2]. Despite aggressive multimodality cancer treatment with chemotherapy, radiation, and surgery, recurrences and premature mortality can occur with a 5-year survival rate for localized disease of 70%^[3] and 33% for those with metastatic

disease^[4].

Pain associated with Ewing sarcoma is the most common symptom reported at diagnosis (88%)^[5]. Its intensity has been described as intermittent and variable with a propensity to become chronic^[6]. Chronic pain in Ewing sarcoma survivors affects approximately 50% of patients^[6]. Nevertheless, there remains a paucity of literature surrounding pain in Ewing sarcoma, particularly its characteristics, severity, associated syndromes, and

effective management strategies^[7].

In this case report, we describe a patient with Ewing sarcoma of the right pelvis which was later complicated by developing secondary chondrosarcoma. This report will highlight the pain characteristics, syndromes, and challenges of pain management of a patient with Ewing sarcoma spanning multiple specialties and exploring key evolving concepts in cancer pain management.

2 CASE REPORT

A 20-year-old man was diagnosed with Ewing sarcoma of the right ilium at age of 9. He underwent neoadjuvant chemotherapy followed by a right hemipelvectomy and then adjuvant pelvic radiation. He had a relapse at age 13 involving the left lung and was treated with chemotherapy, resection of the lung lesion, and radiotherapy. Further relapse occurred at age 19 involving the right lung for which the patient underwent surgery including video-assisted thorascopic surgery further exacerbating his persistent post-surgical pain (See [Figure 1A](#)).

Whilst achieving remission, this patient had developed multiple chronic pain syndromes that significantly impacted his daily activities and function, mobility, school, and work. His chronic pain syndromes included chest pain on a background of pericarditis and bronchiectasis, abdominal pain from a functional gut disorder, and persistent post-surgical pain following his hemipelvectomy.

He required a multimodal and multidisciplinary pain management approach which was led by outpatient chronic pain services. These management strategies included the use of ketamine infusions, opioid rotations, escalation of anti-neuropathic agents, medicinal cannabis, neuroablative interventions, and intensive allied health programs which were also part of his involvement in a chronic pain management program. The pain-related goals included a containment strategy to avoid escalation of analgesia and reduce related side-effects, reduction of his high health care utilization by way of hospitalizations relating to his pain control, and global improvement in function with the associated quality of life benefits.

However, his care remained complicated by his concerns of cancer recurrence. In the context of his chronic pain, the patient noticed increasing right hip discomfort one year following his last treatment. On investigation positron emission tomography scans demonstrated increased avidity in the right hip and an enlarging soft tissue mass which was biopsied, the results of which revealed a dedifferentiated chondrosarcoma. Additional complexity accompanied this secondary cancer diagnosis with the development of a right-sided L5/S1 radiculopathy and an associated foot drop. An MRI revealed tumour progression with invasion into the lumbosacral plexus ([Figure 1B](#)). A right hind-

quarter amputation was recommended and pursued as the definitive management course. This was complicated by wound infection and dehiscence requiring regular dressings. Unfortunately, during his postoperative recovery, the patient was found to have pulmonary metastatic disease, reflected in the escalation and evolution of his previously chronic chest pain ([Figure 1C](#)).

At the time of diagnosis of metastatic disease, the patient was experiencing a combination of acute, chronic, mixed nociceptive, neuropathic, and cancer pain. Acute pain was being experienced in the daily wound dressings, phantom limb pain, and cancer-related pain from the residual tumor invading the lumbosacral plexus, pelvis, and lungs. Given the evolving palliative nature and disease recurrence, palliative care integration was pursued with referral to community services to support care at home, gain adequate symptom control, and optimize quality of life. At this time, the patient required a significant escalation of medications including rotation to long-acting hydromorphone, the use of instant-release fentanyl lozenges, lorazepam, and clonazepam for breakthrough pain. Due to difficulties managing pain, an intrathecal pump had been considered at this time.

3 DISCUSSION

As far as the author's knowledge, there is a paucity of literature reflecting the understanding of the characteristics of pain and effective pain management strategies in those with a diagnosis of Ewing sarcoma^[6]. Though the literature describes the complexity of cancer pain and the potential for experiencing multiple pain syndromes; this case highlights some of the varying and overlapping pain syndromes relating to cancer or cancer treatments ranging from phantom limb pain to persistent post-surgical pain^[6,8]. Ewing sarcoma commonly occurs in the younger population or adolescent period which is a critical time of growth and development with multiple physical and psychological overlays requiring consideration for patient-centered pain management^[6]. In this case, the patient experienced numerous physical and psychological stressors with their pain experience impacting school attendance, functional independence, and the existential distress in considering their own mortality. This case study highlights the importance of appreciating and assessing new onset pain in the context of chronic pain syndromes, particularly in populations at risk of cancer or recurrence of disease.

It also illustrates that those with Ewing sarcoma may experience recalcitrant pain and that the development of chronic pain syndromes over time can be contributed to and exacerbated by multiple external stressors directly and indirectly related to cancer itself. Therefore, it is important to acknowledge and address these issues, when possible, to help with pain management and quality of life.

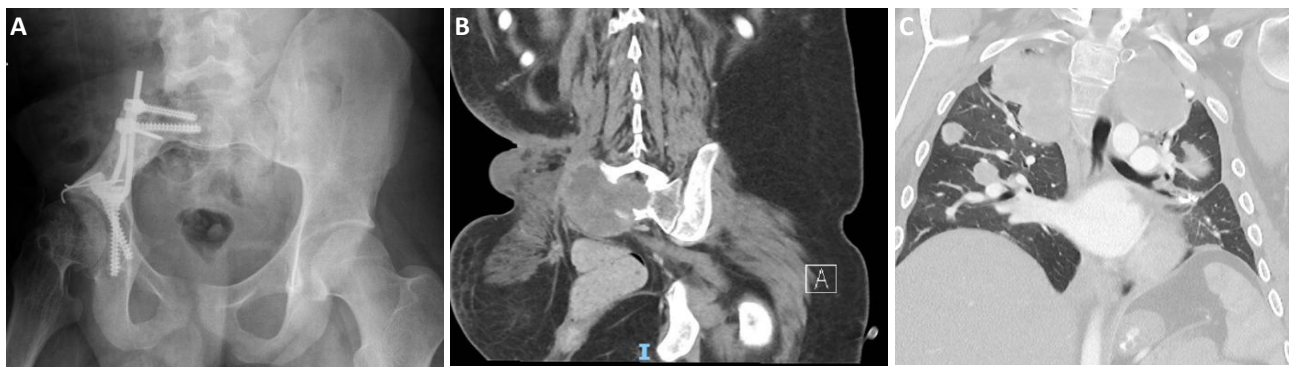


Figure 1. The images of patient conditions contributing to pain. A: X-ray following initial pelvic surgery; B: MRI of right sided hemipelvectomy and cancer progression into pelvis and spine; C: computed tomography of the chest demonstrating progression of pulmonary metastatic disease with cannonball lesions in the lungs.

Table 1. Overview Cancer Pain Management

Phase	Suggested Overview Pain Management
Clinical remission	Suggest multidisciplinary pain management, reduction of medications and polypharmacy.
Stable active disease	Opioids are reasonable but need to work within the opioid stewardship framework.
Disease progression	Symptom-management focus
End of life care	Symptom-management focus

It also highlights the importance and complexity of treating cancer pain where the treatment intent shifts as the disease evolves. This is significant as there are fundamental differences in approaching pain management as the goals of treatment change. In this case, the patient had been managed by chronic non-cancer pain services with a focus on increasing the breadth of pain management strategies to focus on non-pharmacological strategies, rehabilitation, education on sensitization, and opioid reduction^[9,10]. When the patient was diagnosed with a recurrence of cancer and progression of the disease, the escalation of pharmacological approaches including regular and breakthrough opioids reflected the shift to treating nociceptive cancer pain.

During this time, the patient shifted between models of care, demonstrating the importance of clarity and communication between health professionals and health services. The case generates questions about evidence-based best practice when approaching cancer pain, survivorship, and chronic non-cancer pain, recurrence of disease, and progression to supportive palliative care.

It is illustrated within this case study, the need to consider hybrid models of cancer pain management in the context of differing approaches seen in chronic pain and palliative care models^[9]. Perhaps a compromise or middle ground can be achieved between a chronic non-cancer pain model and cancer pain management models. This may be reflected in greater acceptance of pharmacological (including opioid) therapy in appreciating nociceptive and neuropathic mechanisms of cancer pain generation as opposed to the focused non-pharmacological approach and opioid tapering

(or weaning) which is a focus of chronic non-cancer pain programs. Concepts that could be explored in this hybrid model, include embracing ambivalence, education, awareness and acknowledgment of cancer progression and recurrence, weaning of analgesia where possible, including opioids, the use of atypical opioids where appropriate, the use of anti-hyperalgesic agents and strengthening the multidisciplinary approach to pain so that function, quality of life and a sense of wellbeing are maintained for as long as possible (See Table 1).

4 CONCLUSION

This case outlines some of the complexities involved in effectively managing pain in the setting of Ewing sarcoma which includes the predisposing and perpetuating factors for an individual's pain experience. This case was unique in highlighting concomitant sarcoma diagnosis and multiple chronic pain syndromes in a young adult, co-morbidities which had a significant impact on his daily physical and psychological functioning, with an existential overlay in the context of premature mortality. The patient's care involved multiple medical and allied health teams with multimodal analgesic strategies highlighting the significance of pain management in Ewing sarcoma and referral to pain management services critical. Additional research is needed into pain and Ewing sarcoma including prevalence, characteristics of pain, longer-term outcomes in survivors, and interventions / effectiveness of pain management modalities.

Acknowledgements

Not applicable.

Conflicts of Interest

The authors declared no conflict of interest.

Author Contribution

Young J was responsible for writing and the original draft. Lewin J was responsible for reviewing. McGilvray S supervised the overall project. All authors contributed to the manuscript and approved the final version.

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