

Ewing Sarcoma Beyond Adolescence: A Rare Presentation of Spinal Involvement in a 57-Year-Old Male

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1. Abstract

Ewing sarcoma is a rare malignant tumor typically seen in children and young adults, with very few cases reported in individuals over the age of 50. This case describes a 57-year-old male who presented with progressive bilateral hip and back pain, muscle wasting, and limited mobility. Imaging revealed a mass at the L1 vertebral level extending into the paraspinal soft tissues. Biopsy confirmed a diagnosis of Ewing sarcoma, with histology showing small round blue cells and strong CD99 immunoreactivity. Subsequent PET imaging identified a hypermetabolic lesion at L2, suggestive of metastasis. The patient underwent L1 laminectomy and was started on cabozantinib therapy. This case highlights the importance of considering Ewing sarcoma in older adults with atypical spinal or soft tissue lesions. Although rare in this age group, timely diagnosis is critical due to its aggressive nature and poor prognosis in older patients. Increased clinical awareness is essential for early recognition and treatment.

2. Introduction

Ewing sarcoma is an aggressive malignancy primarily affecting bone or soft tissue, and is most commonly diagnosed in children, adolescents, and young adults. It is the second most frequent primary bone cancer in pediatric populations after osteosarcoma, with a characteristic chromosomal translocation, typically t(11;22), resulting in the EWSR1-FLI1 fusion gene.¹ While its peak incidence occurs between ages 10 and 20, cases in older adults, such as 57-year-old men, are exceptionally quite rare. This case report and review of literature examines the incidence of Ewing sarcoma in 57-year-old men, including epidemiological data, age-related trends, gender distribution, and clinical implications, all secondary to a unique patient case.

3. Case Description

The patient is a 57-year-old male with hyperlipidemia on rosuvastatin 10 mg daily, hypertension on metoprolol tartrate 50 mg twice-a-day, and benign prostatic hyperplasia on tamsulosin 0.4 mg daily presenting with progressive bilateral hip pain, back pain and loss of muscle mass for one-month. The patient reports pain with movement and says stretching has not helped. The patient fell and said he had no feeling in his lower body, which caused him to come to the emergency department. There is no history of recent trauma. He rates the pain 8 out of 10, and says that turning, flexing, and bending the lower back and pelvic region causes pain, which have severely affected his activities of daily living. The patient denies any fever, chills, body aches, numbness, or tingling. The patient's blood pressure was elevated at 150/100, but the remainder of his vitals, complete blood count without differential and complete metabolic panel were within normal limits. Physical exam showed mild pain with external

rotation of bilateral hips, and tenderness to palpation at the iliac crests bilaterally, but the remainder of the exam was unremarkable. X-ray of the pelvis with bilateral hips and lumbar spine both showed no acute osseous abnormality. Computed tomography (CT) scan of the abdomen and pelvis shows a large mass at L1 with extension into the paraspinal soft tissues. Interventional radiology performed a biopsy and excision of epidural mass on L1 spine, which showed round cell sarcoma, likely Ewing family (Figure 1-3). CD99 showed strong and diffuse membranous reactivity in the neoplastic cells (figure 4), whereas CK8/18, myogenin, desmin, and CD43 are all negative. The morphology and immunophenotype strongly support a round cell sarcoma, likely a Ewing sarcoma. The patient subsequently underwent L1 laminectomy for resection of Ewing sarcoma epidural mass. The patient later underwent CT positron emission tomography (PET) brow to thigh, which showed a hypermetabolic round focus at the right paraspinal musculature at the level of L2, suspicious for a metastatic implant. He was started on cabozantinib 40 mg daily and continues to follow an oncologist outpatient.

4. Discussion

4.1. Epidemiology, Incidence, and Age-Specific Trends

In the United States, the annual incidence of Ewing sarcoma is approximately 2.9 to 3 cases per million, predominantly affecting individuals under 20 years of age.² It accounts for about 1% of childhood cancers, with a median age at diagnosis of 15 years [1]. The incidence drops drastically beyond the age of 20, with only about 30% of cases occurring in adults over the age of twenty [2]. The number of cases in individuals over 40 are uncommon, comprising less than 10% of total diagnoses [3]. Due to its rarity, there are no explicit reports on the incidence of Ewing sarcoma in adults aged 57. Studies do indicate, however, that the incidence in adults \geq 40 years

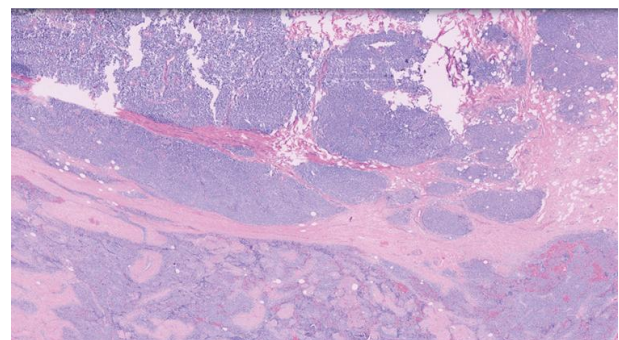


Figure 1: Low power view of small round blue cell tumor.

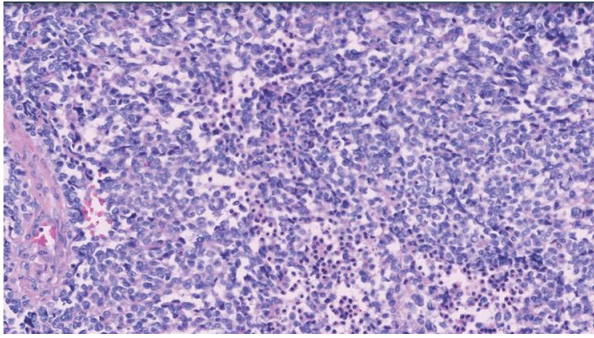


Figure 2: Foci of Characteristic necrosis seen in small round blue cell tumors.

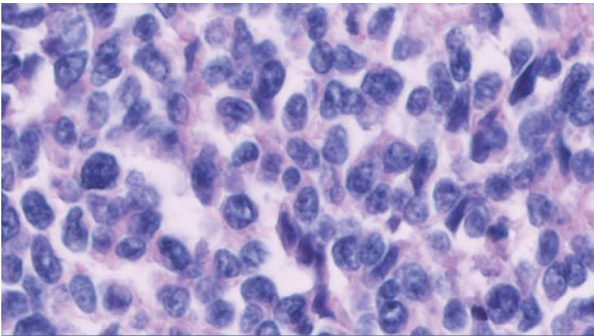


Figure 3: Poorly defined cytoplasm margins and nuclear atypia seen in small round blue cell tumors.

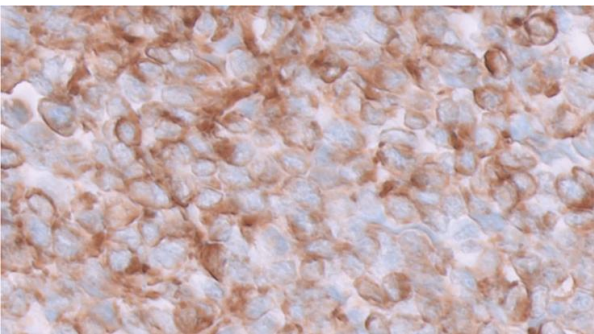


Figure 4: Immunohistochemistry with some membranous staining for CD99, a classic immunohistochemical stain for Ewing sarcoma.

is estimated to be significantly lower than there at less than 0.3 cases per million annually, which is significantly lower than their younger counterparts [4]. More specifically, between 1972 and 2011, there are 2,780 SEER (Surveillance, Epidemiology and End Result cancer program) cases, and only 383 patients (13.8%) were diagnosed at or above age 40, with the proportion decreasing further with advancing age. In addition, the incidence of Ewing sarcoma does show a slight male predominance (male-to-female ratio of approximately 1.3:1 to 1.5:1), a trend which persists amongst all age groups [3]. In addition, Ewing sarcoma is nine to ten times more common in White populations compared to their Black or Asian counterparts, a trend that is consistent amongst all age groups [5]. The peak incidence during adolescence (10–20 years) is likely linked to rapid bone growth during puberty.¹ In older adults, including those around 57 years of age, tumors are more likely to be extraosseous (arising in soft tissue rather than bone), with 66.1% of cases in patients ≥ 40 years being extra-skeletal compared to 31.7% in younger patients [6]. This shift may reflect differences in tumor biology or diagnostic challenges in older populations, where Ewing sarcoma may be mistaken for other sarcomas or metastases.

4.2. Presentation and Immunohistochemistry

Presentation may differ from pediatric cases compared to adults. In younger patients, common sites of involvement include the femur and pelvis, but these may be less frequent in their older counterparts. In older patients, axial tumors (ex: spine, pelvis) or soft tissue primaries predominate.⁶ Presenting symptoms such as localized pain, swelling, or pathological fractures may be attributed to age-related conditions like arthritis or osteoporosis, which can potentially delay diagnosis. Metastatic disease at presentation is also more common in adults (35.5% in those ≥ 40 vs. 30% in younger patients), which could further complicate outcomes in this age group [3]. Although not specific to the disease, Ewing sarcoma typically shows a strong, diffuse membranous expression of CD99, which is a cell surface glycoprotein. EWSR1-FLI1 can also result in expression of the Flt-1 protein, which can further be used to support the diagnosis [7].

4.3. Prognosis and Survival

Survival outcomes in older adults are generally worse compared to children. In younger patients, the five-year survival for localized disease is approximately 68%, but it drops to 39% for metastatic cases [2]. In adults ≥ 40 years of age, survival rates are lower, possibly attributable to a higher likelihood of aggressive disease and reduced tolerance to intensive chemotherapy regimens that are standard in pediatric patients [6].

5. Conclusion

The presentation in our patient of Ewing sarcoma in 57-year-old male is a rare entity, with an incidence likely below 0.1 cases per million annually, far below that of younger populations [4]. The risk is further elevated given the slightly elevated risk given the male predominance and White racial predisposition compared to women or other racial groups. However, the absolute number of cases still remains negligible [5]. The increased prevalence of extraosseous tumors, in addition to higher metastatic rates in older adults compared to younger patients, warrants further investigation [6]. Challenges in studying this population include small sample sizes, potential underreporting, and misdiagnosis, as older patients may be misdiagnosed with more common cancers, or the presentation may be misconstrued for a more common pathology. Future research should focus on further molecular profiling and clinical outcomes in older adults to better understand this subgroup, and increased awareness in physicians as a differential in older patients for atypical cases of bone or soft tissue pathology.

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