A typical Presentation of Ewing Sarcoma in a Pediatric Patient: A Case Report

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Abstract

The Ewing’s sarcoma family of tumors usually occurs in the pediatric population. Although they can arise from any bone or soft tissue, they are most commonly seen in the diaphysis of long bones. We present a case of child who presented with lower extremity weakness and ataxia. Workup revealed a thoracic and lumbar spine mass extending into the neural foramina. Biopsy revealed Ewing’s sarcoma. Urgent surgical decompression was performed which improved neurologic symptoms. The differential diagnosis, work up and management of Ewing’s sarcoma in this atypical location is presented.

Introduction

The Ewing sarcoma family of tumors (ESFT) is a collection of tumors that originate from a common mesenchymal cell type and share similar features, such as histology and chromosomal translocations [1]. Mostly seen in the pediatric population, ESFT commonly arise from the diaphysis of long bones such as the tibia or femur, but can develop in almost all bones or soft tissues [2]. Ewing sarcoma is most common in ages 0-9 years old; however, a higher incidence of Ewing sarcoma in females has been noted at ages 10-14 years old [3]. Only about 8% of all Ewing sarcoma cases originate from the spinal region [4]. Patients with Ewing sarcoma usually present with localized pain that deteriorates rapidly and intensifies at night [5-8]. Constitutional symptoms like fever and weight loss can be seen in 20% of cases; however, they are fairly uncommon [5]. We report an unusual occurrence of spinal Ewing sarcoma that initially presented as gait ataxia, lower back pain, and lower extremity weakness.

Case Presentation

A previously healthy 10-year-old Caucasian female developed left flank and lower back pain over the short course of a month. The pain was initially attributed to the patient’s athletic lifestyle and the patient underwent a trial of non-steroidal anti-inflammatory drugs, heat and rest. The pain progressively became worse and started waking her up at night. The patient finally presented to the pediatric emergency room, one month after initial onset of symptoms, with severe aching pain, balance problems, lower extremity weakness, and gait ataxia. The pain was localized to the left lower back and flank area; it was exacerbated by lying flat. The patient also complained of heavy legs, difficulty walking, difficulty balancing, and paresthesia of the feet. On exam, cranial nerves two through twelve were intact bilaterally. Sensation to touch was present in all extremities including her legs. Decreased rectal sphincter tone was noted on exam. Upper extremity muscle strength was MRC grade 5/5. Lower extremity examination revealed decreased range of motion, decreased reflexes, and decreased muscle strength. Pertinent lab work showed a slightly elevated LDH at 313 U/L. Chest radiograph showed a 1.5 cm nodular density at the right lung base, moderate vertebral body height loss at T9, and minimal loss of T8 height [Figure 1 A and 1B]. MRI of the cervical, thoracic, and lumbar spine as well as a brain MRI were performed. Diffuse multifocal hypo-intensities and epidural masses were seen within the thoracic and cervical spines as well as multiple lesions in the vertebral bodies on T1 weighted images [Figure 2A and 2B]. Additionally, a soft tissue lesion was seen in the neural foramen of L1-L2 [Figure 3]. The epidural mass extended into the right neural foramen causing expansion of the foramen. Biopsy of one of the spinal epidural mass was performed. This showed a small round blue cell tumor, consistent with Ewing sarcoma/peripheral neuroectodermal tumor, infiltrating the fibroadipose tissue. CT axial imaging was performed and demonstrated pulmonary metastatic lesions, primarily in the right lower lobe. PET scan showed hypermetabolic pulmonary nodules as well as diffusely increased metabolic activity throughout the bone marrow. Because of her rapidly declining neurological status, the decision was made to take the patient urgently from MRI to the operating room for spinal cord decompression and tumor removal. Post-operatively, the patient’s pain was much improved and some function of her lower extremities returned. Intensive physical therapy was started a few days after surgery in order to improve the patient’s strength and coordination. Chemotherapy was also started after surgery. It was felt by the patient’s specialists that decompressing the spinal cord took precedence over beginning her chemotherapy.
Figure 1(A,B): Chest X-ray showing a nodular density at the right lung base (yellow circle) (A) Anterior Posterior View (B) Lateral View

Figure 2(A,B): MRI of the T-Spine showing a lesion in the right neural foramen at T7-T8. (A) T2 Weighted T-Spine MRI, Axial View; (B) T1 Weighted T-Spine MRI, Sagittal View

Figure 3: T2 weighted MRI of the lumbar spine showing a soft tissue lesion in the neural foramen of L1-L2
Discussion

Ewing sarcoma typically occurs in the extremities, specifically in long proximal bones [9]. Primary lesions are most commonly seen in the diaphysis of the femur, tibia, and humerus [2, 4]. Although the proximal long bones are the most common primary location site, they can originate from any bone or soft tissue [2, 4] of the 54% of EFTs that originate from the axial skeleton; roughly 8% of those arise from the spine [4]. Unfortunately, our patient presented with one of the rarer forms of Ewing sarcoma. Her sarcoma was found to have originated in the spine, which typically spells a worse prognosis for the patient. A number of characteristics can help define the prognosis, as well as direct the optimal modality of treatment. These prognostic factors include size and location of tumor, presence metastases, patient age, and presence of certain chromosomal translocations [10]. Patients presenting with axial primary tumors e.g. in the pelvis, rib, spine, scapula, skull, clavicle, or sternum tend to have a worse treatment outcome than those who present with extremity lesions such as the femur or tibia [4, 11]. One study showed that the five-year relapse-free survival rate of tumors in the extremities versus axial tumors was 40 versus 61 percent [4]. In addition, patients with smaller primary tumors tend to do better than those with larger tumors [12]. Fever, anemia, and elevated serum lactate dehydrogenase (LDH) correlate with a greater burden of disease and therefore worse prognosis [4, 13]. A short interval between onset of symptoms and diagnosis is likely to be associated with metastatic disease [13, 14]. This poorer prognosis of spinal primary lesions is likely due to the difficulty of achieving negative resection margins. Radical resections or removing the entire epidural space may not be feasible. A retrospective study of roughly 1,000 patients approximated five-year survival rates for patients with localized disease at 55% and 21% for patients that present with metastasis at the time of diagnosis [4]. Another retrospective study of 300 patients, showed a 65.2% five-year survival rate overall. It also showed a 78.6% five-year survival rate for patients with localized disease and a 28.1% five-year survival rate for patients with extra-pulmonary metastasis [15]. Unfortunately, our patient presented with primary lesions in the spinal cord, as well as widespread lung metastasis.

It is important to consider other etiologies for our patient’s presenting symptoms during the initial work-up. Our patient primarily presented with gait ataxia, lower extremity weakness, balance problems, and pain. The patient also noted paresthesia’s and had decreased rectal tone on examination. Transverse myelitis was considered as a possible etiology for her symptoms, however the patients back was mostly unilateral and she didn’t endorse sensory issues [16]. Guillain-Barre was also considered due to her inability to walk and her balance issues, but ruled out by her rectal sphincter dysfunction (pointed more towards spinal cord compression as opposed to an inflammatory demyelination), imaging, and lack of a preceding infection [17]. Her coordination problems could have been attributed to a first attack of multiple sclerosis, but imaging pointed away from this possibility [18]. The mix of her symptoms alongside imaging pointed towards some sort of pediatric malignancy. A neuroblastoma pressing on the spinal cord can cause weakness, an inability to walk or stand, and areflexia or hyporeflexia as seen in our patient [19]. The majority of neuroblastoma cases are sporadic which could explain the patient’s lack of familial history. Another form of malignancy that was considered was a rhabdomyosarcoma; rhabdomyosarcomas usually present as a mass with signs and symptoms that vary based on location. A common metastatic site is the lungs, which was present in our patient [20]. Finally, a diagnosis of medulloblastoma was possible in our patient due to extreme spinal cord involvement. However, patients usually present with increased intracranial pressure secondary to 4th ventricle blockage. The blockage contributes to neural symptoms such as vomiting, dizziness, diplopia, but none of these symptoms were noted in our patient [21]. In the end, the presenting symptoms along with imaging and pathology pointed towards Ewing sarcoma as the final diagnosis. After surgical decompression of the thoracic spine, the patient started to regain function in her lower extremities. The patient’s control of her legs started to improve significantly as the days passed post-op; this pointed towards an etiology of secondary ataxia and lower extremity weakness caused by spinal cord compression from the primary Ewing sarcoma insult. Her balance improved significantly and she was eventually able to stand and ambulate without parental support. She is currently undergoing physical therapy in order to improve lower extremity function. However, based upon the widespread disease at presentation, her prognosis is not favorable.

References

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