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Case Report

Sacrococcygeal Ewing Sarcoma: A Case Report

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Abstract

In this report, we present a rare case of Ewing's sarcoma with a peripheral primitive neuroectodermal tumor (ES/PNET) arising from presacral soft tissue in a 14-year-old boy. He had a previous long history for 2 years ago: dramatically started by progressive, worsening low back pain three months of trauma; to ending with a paraplegia associated with urine and stool incontinence. He was presented to a hospital in the capital Sana'a, Yemen. Computed tomography (CT) scan and magnetic resonance imaging (MRI) showed a soft tissue mass in presacral area with bone erosion and destruction; staging chest radiographs and CT showed no evidence of metastatic disease. While, his histologic testing confirmed presence of " ES " as small, round, blue cells. The patient subjected to chemotherapy for one year followed by radiotherapy and he got improved. He stopped radiotherapy for four months led to rapid progressive deterioration ended by brain metastasis and the patient died through three months.

Keywords: Ewing's sarcoma, Presacral soft tissue, Case report, Yemen

1. Introduction

Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET) is a highly malignant neoplasm which belongs to group of closely related tumors share some common features and called typically Ewing sarcoma family of tumors that commonly arise from the bones and rarely in soft tissue. These groups of small, blue, round cell neoplasm include the well-known Ewing sarcoma (EWS), PNET, and Askin tumor [1]. Ewing's sarcoma (ES) has unknown cause. The chromosomal translocation t(11;22)(q24;q12) is present in more than 85% of cases that diagnosed as ES [2].

ES occurs in a wide range of ages from infants to the elderly [3], generally affects white population [4], and has a predilection for the male sex (male/female ratio, 1.3-1.5:1) [3,4]. We reported here a rare case presented by unusual presentation (paraplegia: stool and urine incontinence) diagnosed as presacral soft tissue Ewing sarcoma.

2. Case report

A 14-year-old boy, nonsmoker, with low socioeconomic level from Yemen is diagnosed with sacrococcygeal Ewing sarcoma. The case started in more than two years ago with a history of progressive, worsening low back pain which was burning in nature associated with weakness and tingling sensation in both legs. These symptoms and signs happened three months after trauma (accident). The case couldn't be controlled by analgesic and his condition complicated through six months to end by paraplegia associated with urine and stool incontinence. So, the patient was taken to a hospital in Sana`a for investigations. CT and MRI showed a soft tissue mass in presacral area with bone erosion and destruction (Figure 1).

The case showed no evidence of metastatic disease at presentation as determined by staging chest radiographs and CT (Figures 2,3). He had histologic confirmation of " ES " small, round, blue cells showed in histopathologic reports and immunohistochemistry. The patient

subjected to chemotherapy for one year followed by radiotherapy. When he got improved with tumor regressed as reported in MRI (Figure 4), and has started to walk again, but stopped radiotherapy for four months. So, he relapsed and his condition worsened and developed paraplegia again associated with legs muscles wasting (Figure 5).

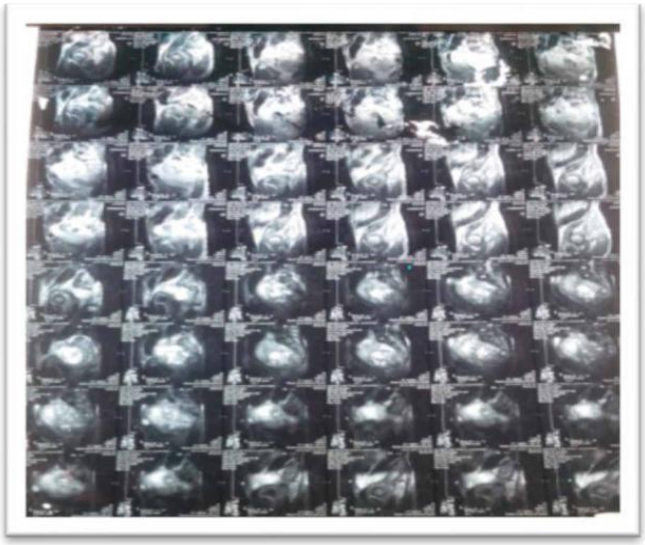

Figure 1: CT of the pelvis shows tissue mass in presacral area

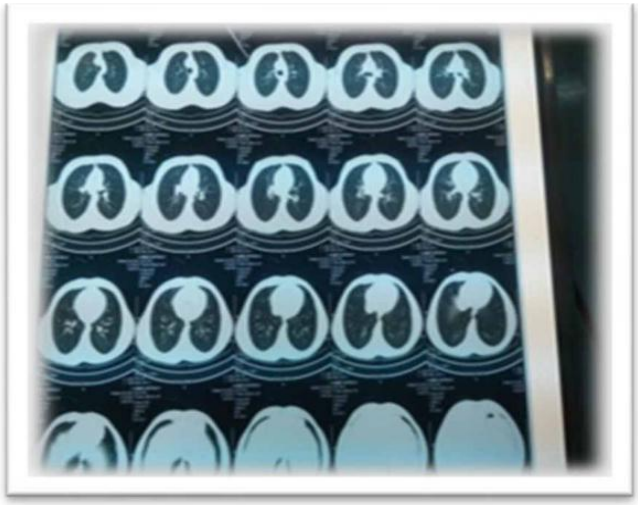

Figure 2: CT of the chest shows no lung metastasis

He developed bed sore due to prolong immobility (Figure 6), then he started to develop a brain mass increased with the time through two months (Figure 7) also he suffered chronic constipation. So, he returned to radiotherapy with continuous using of laxative. Consequently, he was taken to hospital where he subjected to laparotomy and colostomy operation, then he was shifted to ICU. The patient's condition continued to deteriorate and he died.

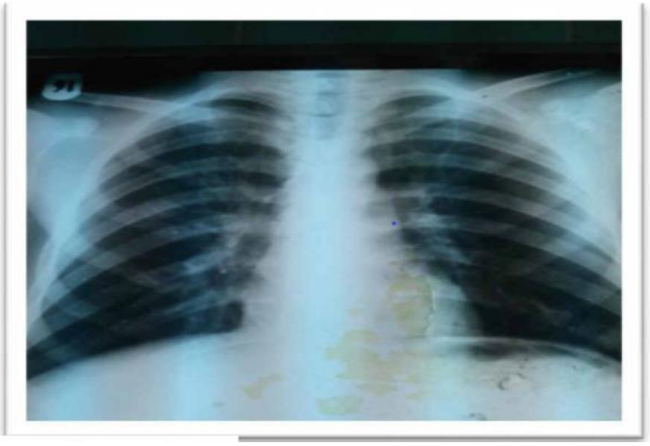

Figure 3: Chest radiograph shows normal chest appearance

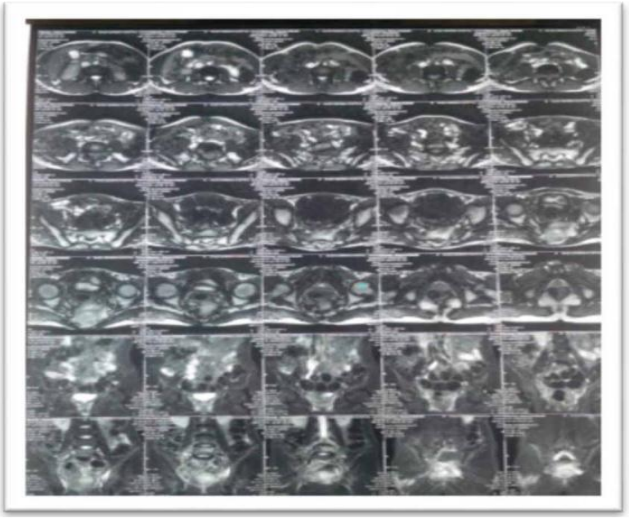

Figure 4: MRI post radiotherapy

Figure 5: Patient with lower limb weakness

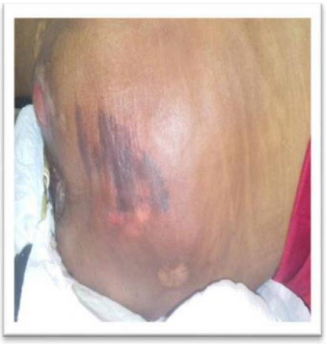

Figure 6: Patient with bed sore

Figure 7: Patient with cranial metastasis

3. Discussion

The ES of presacral lesions are rare [5]. Ewing sarcoma occur in about 1 % of all childhood cancers [6]. In the family of PNET/ ES, they are part of rare group of nonhereditary malignant tumors [7] which are generally originating in bone, but they can sometimes originate in soft tissue, known as extraosseous and predominantly observed in adolescents and young adult between 10-30 of age [3]. It is considered as aggressive course and high relapse rate [7]. Extraosseous ES/PNET occur mainly at paravertebral region, lower limbs and chest wall and less commonly in the pelvic cavity, retroperitoneal region or the upper limbs [8], head and neck [9].

Our patient has presacral soft tissue ES diagnosed by pelvic CT with differential diagnosis was sacral chordoma, sacrococcygeal teratoma [10]. Hence, it is confirmed by histopathology and histoimmunochemistry workup [6]. The main clinical presentation of this disease is deep soft tissue mass that not show redness, swelling and other inflammatory signs in the surface and, local pain may occur. The tumors that are near the spine rarely can cause weakness, numbness, or paraplegia in the legs [11]. Our patient presented by low back pain associated with paraplegia and, urine & stool incontinence. This is a rare presentation. Almost the first treatment of ES is the chemotherapy as systemic control in case of micro-metastasis [12] that can't appear by imaging [11] followed by localized therapy as surgery and/or radiotherapy [13].

Once the ES has been diagnosed and staged chemotherapy started, this is called neoadjuvant chemotherapy. It is given before any surgery or radiation therapy. So, imaging tests are done at least 12 weeks post chemotherapy to see if the tumor is shrinking and can be surgically removed [3]. Localized tumors, smaller tumor size, and primary tumor in arms or legs (opposite to chest or pelvis), normal LDH and, age younger than 10 years in addition to good response to chemotherapy have a better prognosis [11]. Our patient was diagnosed initially as ES with no metastasis appeared, he responded to chemotherapy and started to improve. The tumor followed unusual course after our patient was neglected at the end of the course of treatment when he stopped radiotherapy. Therefore, he showed rapid progressive deterioration ended by brain metastasis and he died after three months.

4. Conclusion

Although, the ES/PNET in the soft tissue is very rare. It can mimic to a great extent some malignant neoplasm. It demands early detection followed by early investigations using radiography, CT, and MRI followed by histopathology and immunohistochemistry. These investigations are necessary for early accurate diagnosis that is vital for the patient. The tumor presents at a younger age and is very aggressive. It usually has a poor prognosis. Here the patient has been died in spite of treatment by chemotherapy and radiotherapy.

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Competing interests
The authors declare that they have no competing interests.

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