AKUT KARINDA NADİR BİR NEDEN: PRİMER İSKELET DIŞI EWING SARKOMU

A Rare Cause of Acute Abdomen: Primary Extraskeletal Ewing Sarcoma

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ÖZET

Yirmi altı yaşında erkek hasta akut karın ile başvurdu. Parasentezde karın içi kanama görüldü. Eksplorasyonda rektosigmoid kolonu ve mesaneyi invaze eden pelvik kitle saptandı. Doku biyopsi incelemesinde, vimentin ve CD99 için pozitif boyanmış Ewing sarkom tumor hücreleri gösterildi. Bu vakada, pelvisin sıradışı bir tümörü olan iskelet dışı Ewing sarkomunu sunduk.

Anahtar kelimeler: *Akut batın; Ewing sarkomu; İskelet dışı*

ABSTRACT

A 26-year old man was presented with an acute abdomen. Paracentesis revealed hemoperitoneum and a pelvic mass invaded to rectosigmoid colon and bladder was detected during exploration. Tissue biopsy examination demonstrated Vimentin and CD 99 positive stained tumor cells of the Ewing sarcoma. In the present case, we have reported an unusual tumor of the soft tissue of the pelvis, an extraskeletal Ewing sarcoma.

Keywords: *Acute abdomen; Ewing sarcoma; Extraskeletal*

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INTRODUCTION

Ewing sarcoma (ES) was first defined in 1921 by James Ewing as a malignant small round cell bone tumor in a 14-year-old girl. ES is a distinct round-cell sarcoma that occurs predominantly in the long bones of skeletally immature patients. The tumor is composed of undifferentiated, round, mesenchymal cells that are rich in glycogen and typically manifest a unique reciprocal chromosomal translocation, t(11;22)(q24;q12), which occurs in approximately 90% of these tumors. Very few other human tumors exhibit such consistent karyotypic alterations, which might play a significant role in their pathogenesis. Extraskeletal ES is a rare soft tissue tumor. "Ewing Sarcoma Family of Tumors" consists of peripheral primitive neuroectodermal tumor (PNET), extraskeletal ES, and Askin Tumor (ES of the chest wall) as well as ES (1). The aim of the present case report is to emphasize the importance of planning the surgical strategy due to either non-frequent occurrence or very rare leading to acute abdomen with spontaneous perforation of ES.

CASE REPORT

A 26-year-old male patient applied to emergency department due to acute abdominal pain. Physical examination showed unremarkable abdominal sounds, rigid and tender abdomen with rebound. Abdominal paracentesis revealed hemoperitoneum and therefore exploratory laparotomy was performed without further investigation. A hemorrhagic unresectable mass involving complete pelvis, bladder and rectosigmoid colon was detected during the exploration. With these findings, the patient was referred to our clinic with placing the drains in the abdominal cavity for further investigation and treatment. Abdominal computed tomography (CT) showed a 14x11x14 cm retrovesically placed mass (Figure 1). Fatty plains were absent between the pelvic mass and bladder. Thorax CT of the patient was unremarkable. No extra focuses were detected in whole body bone scintigraphy. In the colonoscopy, extramucosal mass bumped in the anterior wall of the rectum was observed. Oral feeding was started to the patient who was given antibiotic (ceftriaxone + metronidazole) and IV fluid and electrolytes. Prostate Specific Antigen values and tumor markers (CEA, AFP, Ca 15-3, Ca 19-9) of the patients were found within normal limits. Histopathologic examination of the biopsy material using hematoxylin and eosin revealed small round tumor cells (Figure 2). Immunohistochemical study showed positively staining Vimentin and CD99, focal and weak staining neuron-specific enolase (NSE) and synaptophysin, and negatively staining pancytokeratin, chromogranin, lymphocyte common antigen (LCA), Desmin, CD117, CD34, Tolt, epithelial membrane antigen (EMA), myogenin, CD10, placental-like alkaline phosphatase (PLAP) and beta human chorionic gonadotrophin (BHCG) (Figure 2). Rare intracytoplasmic granules were observed in the PAS staining. Ki-67 proliferation index was found as 10%. The lesion diagnosed as Ewing's sarcoma/primitive neuroectodermal tumor (PNET/ES). The patient was found eligible for chemotherapy in the evaluation of the oncology council. He was started to the treatment of adriamycin, cyclophosphamide and vincristine.

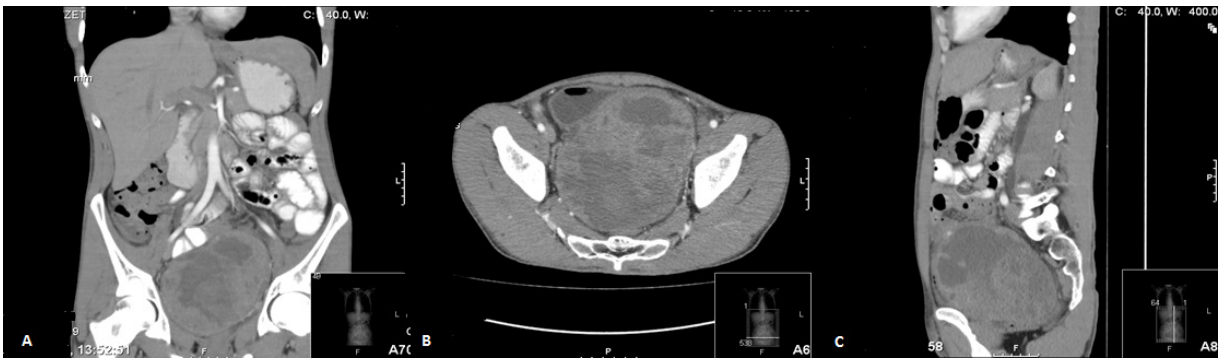

Figure 1. A. Coronal section of abdominal CT showed a 14x11x14 cm retrovesically placed mass. B. CT section showed the same lesion in axial view. C. Sagittal section of abdominal CT showed the retrovesically placed mass which fills the pelvis.

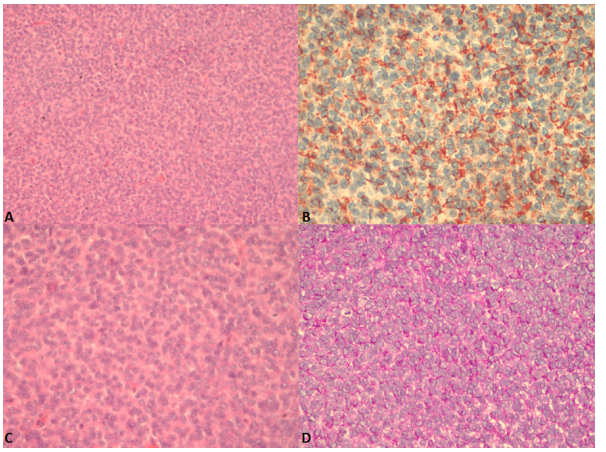

Figure 2. A. Small round tumor cells (Hematoxylin and eosin, x200). B. Immunohistochemical study showed positively staining CD99 (x400). C. Demonstration of small round tumor cells in high magnification (Hematoxylin and eosin, x400). D. PAS positive intracytoplasmic granules were observed (x400).

DISCUSSION

Sarcomas are unusual but not rare malignancies. They account for only 1% of adult solid tumors. These sarcomas appear most frequently between the fourth and sixth decades of life with a 2:1 male/female ratio. They can arise anywhere in the body with the lower extremity being the most common site. Incidence are as follows: lower extremities (46%), upper extremities (13%), retroperitoneum, pelvis and visceral (12%), truncal sarcomas (19%), and head and neck sarcomas (9%) (2). The presenting symptoms and signs of all sarcomas are usually nonspecific. With sarcomas involving the abdominal and pelvic cavity diagnosis is even more subtle because these tumors may progress for long time periods without causing overt symptoms (1). In this present case report, we defined a ES in 26-year old male which was presented as acute abdomen.

Ewing's sarcoma is the third most common primary bone sarcoma. It has a significant predilection for the white population. The peak incidence is the second decade of life. In very young patients, and in patients over the age of 30, a diagnosis of Ewing's sarcoma should be questioned, because it occurs so rarely in these age groups. In extraskeletal ES, the male/female rate is reported as 1.5:1 in contrast to osseous form that is specific to a gender. The age range that the tumor is commonly observed is reported between 4-47 years. Despite the tumors occur as a mass without pain in patients with extremity lesions, there may be pains sourced from other body sites. It is reported that extraskeletal ES can originate from various body sites including scalp, larynx, nasal fossa, neck, chest wall, lungs, paravertebral soft tissues, pelvis, perineum, arms, legs, fingers, and toes (3). However, the most common sites that these tumors can be seen are reported to be paravertebral soft tissues and lower extremities.

Extraskeletal ES can be diagnosed basically depending on the characteristics of the tumor detected via light microscopy, and so it can be differentiated from round cell soft tissue tumors, malign lymphoma, and hemangioperistoma in many cases. A case of ES originated from kidney was diagnosed after spontaneous rupture of the lesion was reported (4). In our case pathologic examination of biopsy material revealed small round tumor cells. After detailed immunohistochemical examination for differential diagnosis, the lesion firstly was thought as PNET/ES.

CONCLUSION

In conclusion, it is reported that extraskeletal ES can originate from various body sites. In this case report, we defined a very rare intraperitoneal extraskeletal ES which presented with acute abdomen. Extraskeletal ES should be kept in mind in the differential diagnosis of pelvis soft tissue tumors particularly in young patients even if it is very rare.

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