Case Report

Primary Extraskeletal Ewing's Sarcoma Presenting as an Axillary Mass: A Case Report and Review of the Literature

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Abstract

Extraskeletal Ewing's sarcoma (EES) is a rare soft tissue tumor predominantly observed in adolescents and young adults, and is characterized by aggressive behavior. So far, only two cases of primary axillary soft tissue EES have been reported in the literature. One of them was a 29-year-old female patient who presented with a lump in her left axilla. Upon examination, an irregular, painless mass, measuring 5 cm × 5 cm × 3 cm, was noted in the left axilla. A histopathological examination of the mass revealed small, round, blue cells with scant cytoplasm, round nuclei, numerous mitosis, and necrosis. An immunohistochemistry (IHC) examination was positive for CD99 and negative for ER, PR, Her2neu, CK7, CK5/6, CD56, CD45, CK-pan, CKHMW, P63, desmin, S100, TdT, vimentin, myogenin, synaptophysin, and chromogranin A. The patient was diagnosed with primary axillary soft tissue EES and was started on neoadjuvant chemotherapy. Twelve months later, she is clinically free from the disease.

Keywords: *Axilla, extraskeletal Ewing's sarcoma, Madinah, Saudi Arabia*

Introduction

Ewing sarcoma (ES) is a rare type of cancer that usually affects children and young adults. Males are affected more often than females. It usually arises in the long bones of the lower limbs, and the bones of the pelvis and chest wall are also often affected.[1] Extraskeletal Ewing's sarcoma (EES) is a rare soft tissue tumor predominantly observed in young patients between the ages of 10 and 30, and it is characterized by aggressive behavior with a high death rate.[2] Most commonly, it arises in the soft tissue of the paravertebral region, the lower extremities, the chest wall, and the retroperitoneum, but it has also been reported in various sites such as the breast, pancreas, lungs, biliary tract, kidney, prostate, stomach, esophagus, oral cavity, salivary glands, urinary bladder, uterus, cervix, gonads, and vagina.[3] The axillary location of EES appears to be very rare; so far, only two cases have been reported in the literature.[4,5] The objective of the present case report is to document a rare case of EES originating in the axillary region in a young Saudi female from the Madinah region in Saudi Arabia, and to highlight the importance of immunohistochemistry (IHC) in the differential diagnosis of this rare tumor at an unusual site.

Case Report

Patient information

An otherwise healthy 29-year-old Saudi female patient presented at the Breast Clinic of King Fahad Hospital in Madinah, Saudi Arabia, with a lump in her left axilla that had been present for the last eight weeks. She had no similar swelling elsewhere in the body and no history of any malignancy in the past.

Clinical findings

On examination, an irregular, painless mass, measuring 5 cm × 5 cm × 3 cm, was noted in the left axilla. Both the breasts and the contralateral axillary area

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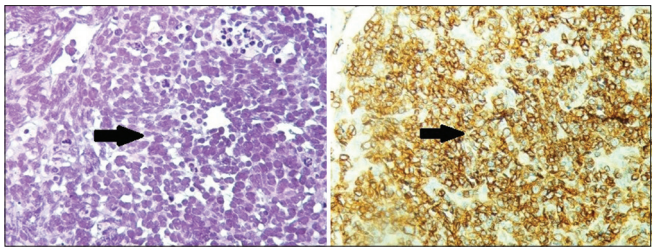

Figure 1: Microphotography showing small round blue cells with scant amounts of ill-defined cytoplasm, atypical mitosis (hematoxylin-eosin at Magnification × 40), and positive staining of tumor cells for CD99 (Avidin Biotin at magnification × 40). (Arrows indicate malignant cells)

were clinically normal. Systemic examination was unremarkable.

Diagnostic assessment

Ultrasonography revealed a large, irregular lobulated heterogeneous hypoechoic mass measuring approximately 4.7 cm × 4.5 cm in the left axilla with benign-looking left axillary lymph nodes and left breast cysts. The patient underwent an ultrasound-guided Tru-Cut needle biopsy, and the specimen was sent to the Pathology Department at King Fahad Hospital. The microscopic examination revealed multiple fragments of fibrocellular tissue showing irregular lobules and clusters of malignant cells arranged in organoid and filigree patterns. These lobules and clusters were separated by fibrous strands consisting of monomorphic small round blue cells with scant cytoplasm, round hyperchromatic nuclei, numerous mitosis, and necrosis. No breast or lymphoid tissue was seen [Figure 1]. A provisional diagnosis of a small round blue cell tumor was made, and a wide range of IHC panels were run to confirm the diagnosis. The tumor cells were strongly positive for CD99 and negative for estrogen receptor (ER), progesterone receptor (PR), Her2neu, cytokeratin 7 (CK7), cytokeratin 5/6 (CK5/6), CD56, CD45, pan-cytokeratin (CK-pan), CKHMW, tumor protein 63 (p63), desmin, S100, TdT, vimentin, myogenin, synaptophysin, and chromogranin A [Figure 1]. Subsequent computed tomographic (CT) scans of the chest, abdomen, and pelvis along with bone scans revealed no evidence of an occult primary lesion. The morphological features and IHC profile were compatible with primary axillary soft tissue EES.

Therapeutic intervention

A month later after presentation, the patient underwent a wide local excision. Subsequently, she was administered adjuvant chemotherapy with vincristine, cyclophosphamide, and doxorubicin (3-week intervals between cycles). Local radiation therapy of the axilla was performed after 6 cycles of chemotherapy.

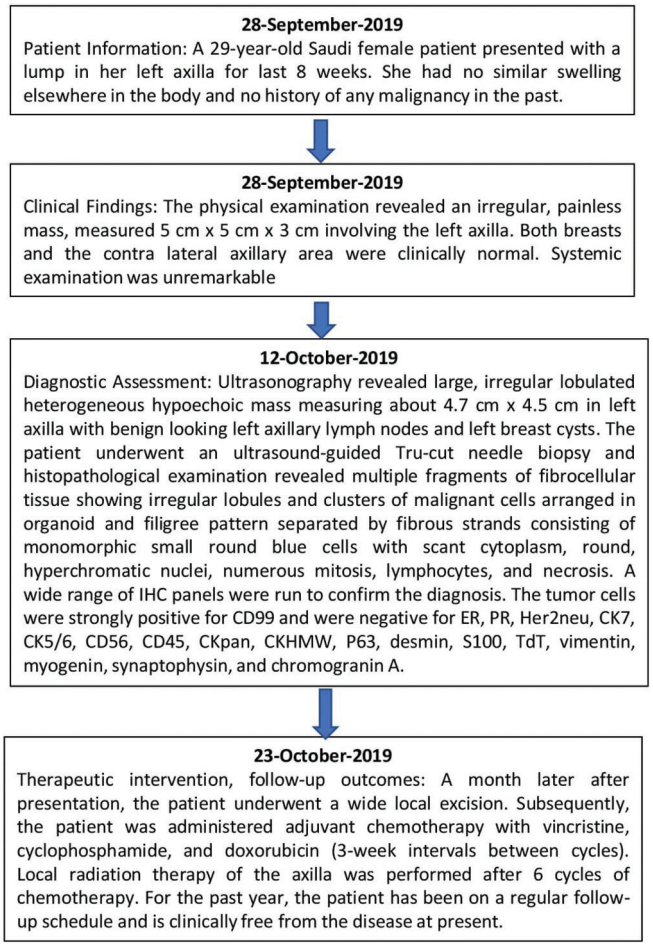

Figure 2: Timeline summary of the patient's current illness, clinical examinations, diagnostic evaluations, therapeutic management and follow-up.

Follow-up and outcomes

For the past year, the patient has been on a regular follow-up schedule and is clinically free from the disease at present [Figure 2]. The case report was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Informed consent was not obtained from the patient due to no communication.

Discussion

Extraskeletal Ewing's sarcoma was first described by Tefft *et al.*[6] in 1969, who reported four patients with paravertebral soft tissue tumors that were histologically similar to primary ES of bone. In 1975, Angervall and Enzinger reviewed the pathologic features and behavior of 39 EESs, occurring predominantly in young adults. The clinical course is rapidly progressive with a high death rate. The most common sites are the paravertebral region and lower extremities, followed by the chest wall, retroperitoneum, pelvic and hip region, and upper extremities.[7]

An extensive literature search revealed only two cases in which the diagnosis of primary axillary soft tissue EES

was reported. The first one, reported by Murphey *et al.*[4] in 2013, described EES in an 11-year-old boy with an enlarging axillary soft tissue mass in his left axilla. The second case was reported by Chatterjee *et al.*[5] in 2017 when they reviewed 50 adult patients with the ES family of tumors (ESFT). Our case is thus the third reported case of this tumor at this uncommon anatomical site, which indicates the rarity of this lesion. Occurrence at unusual sites can lead to consideration of alternative clinical diagnoses and make the evaluation process more challenging.

Several case reports on primary EES showed that patients were mainly young, between the ages of 20 and 35 years,[2,3]which was also the case with our patient. However, Gazula *et al.*[8]reported a primary EES, in a four-month-old girl, masquerading as an infantile benign neck mass, and Charlotte *et al.*[9]reported an 80-year-old woman with intra-abdominal EES. To the best of our knowledge, these are the youngest and oldest reported cases, respectively, so far.

Immunophenotyping and cytogenetic study are necessary to confirm the diagnosis of ES. The positive expression of CD99 (MIC2), a cell surface glycoprotein involved in cell adhesion, plays a crucial role in the diagnosis of ES, as it is expressed in more than 90% of cases.[8] However, CD99 may also be expressed in other tumors, including malignant lymphoma, neuroendocrine carcinoma, rhabdomyosarcoma, small cell carcinoma, and neuroblastoma.[9] In our patient, malignant lymphoma, neuroendocrine carcinoma, rhabdomyosarcoma, small cell carcinoma, and neuroblastoma were excluded by negative staining for cytokeratins, TdT, CD45, CD56, S100, desmin, myogenin, and chromogranin A. The translocation t (11, 22) (q24; q12) is specific to ES and results in the formation of a chimeric fusion transcript between the EWS gene on chromosome 22 and FLI-1 chromosome 11, and this translocation is found in more than 90% of cases.[10] However, we were unable to conduct this genetic test due to the lack of translocation assay kits in our genetic laboratory at the time.

Conclusion

To conclude, a primary ES arising from axillary soft tissue is exceedingly rare, and only two cases have been reported in the literature (English) so far. This case highlights the rarity of this lesion and the importance of IHC examination in the differential diagnosis of this rare tumor and its occurrence at an unusual site.

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Conflicts of interest

There are no conflicts of interest.

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