Cutaneous Ewing's Sarcoma of Scalp with Lung Metastases in an Elderly Female: A Rare Case Report

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Abstract

Ewing's sarcoma (ES) is a primitive neuroectodermal tumor. It is usually a primary bone tumor but rarely occurs in the skin and subcutaneous tissues (primary cutaneous ES [PCES]). It usually involves the deep subcutaneous tissue or muscles and rarely occurs as primary skin cancer. Most patients are white, women, and in the second decade of life. The diagnosis is made by aspiration cytology, histochemical stains, immunohistochemistry, electron microscopy, cytogenetics, and molecular genetics of translocations. Due to their rarity and morphological similarity to other cutaneous tumors, ESs are subject to being clinically and pathologically underdiagnosed. Cutaneous ES has a better prognosis than primary bone or soft-tissue ES, with a survival rate of 91% in 10 years and the presence of metastasis being rare. At present, no specific treatment guidelines inform the management of cutaneous Ewing tumor. The treatment modalities are extrapolated based on the management of bone ES while includes neoadjuvant chemotherapy, surgery, adjuvant chemotherapy (±radiotherapy), and autologous bone marrow transplantation in high-risk patients. Standard first-line treatment for patients with these tumors includes chemotherapy with a five-drug regimen of vincristine, doxorubicin (Adriamycin®), and cyclophosphamide, alternating with ifosfamide and etoposide. We report a rare case in a 60-year-old female diagnosed as PCES with lung metastases, treated by palliative chemotherapy.

Keywords: *Ewing's sarcoma, palliative chemotherapy, primary cutaneous Ewing's sarcoma, vincristine, doxorubicin (Adriamycin®), and cyclophosphamide regimen*

Introduction

Primary cutaneous Ewing's sarcoma (PCES) is a very rare tumor with very few cases being reported worldwide.[1,2] Only 78 cases had been described in the literature up to 2001, with the majority of patients in the second decade of life.[1,3-5] PCES is a malignant type of skin tumor that affects older children. Ewing's family of tumors include Ewing's sarcoma (ES) of bone, extraosseous Ewing's tumor, primitive neuroectodermal tumor (PNET), including medulloblastoma and Askin's tumor.[6] This classification allows rare tumor such as PNETs to benefit from chemotherapy regimens traditionally reserved for skeletal ES.[4] Location of this tumor in decreasing order of frequency is the lower limbs, upper limbs, head and neck, followed by the trunk. The tumor presents as a mass under the skin, usually 2–3 cm in size, which is painless. Any combination of chemotherapy, radiation therapy, and invasive procedures are used to treat this tumor. The overall survival rate with this tumor type is around 93%. Better prognosis is associated with diagnosis at an early stage, a small-sized tumor, and absence of metastasis.[1-3,5,7] Around 65% of patients survive in localized PCES as compared to less than one-third survival in metastatic patients. Improved outcomes have been obtained with radiotherapy and chemotherapy and the survival rate is above 40%.[1,3,4]

Case Report

A 60-year-old female with no past medical history, no addiction history presented with progressively increasing swelling over the left side of scalp since the last 12 months, with no associated pain, fever, and history of weight loss. On local examination, there was an 8 cm × 10 cm slightly foul-smelling, cauliflower-fungating growth over the left side of the scalp. Contrast-enhanced computed tomography (CECT) of the brain, neck, thorax, and abdomen revealed evidence

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of a large moderately enhancing lesion measuring 3.4 cm × 10.9 cm × 9.5 cm (TR (Transverse section) × AP (Anteroposterior) × CC (Craniocaudal section)), involving the scalp overlying left frontoparietal temporal region with underlying bony remodeling without intracranial extension. Furthermore, there was evidence of heterogeneously enhancing lesion in the left apical region [Figure 1]. A surgical biopsy was suggestive of small blue round cell tumor showing round-to-oval nuclei, vesicular nuclei, small nucleoli, and thin eosinophilic cytoplasm. On immunohistochemistry (IHC), cells were immunoreactive for vimentin, CD99, FLI-1 and negative for CK, LCA, S100, HMB-45, melan-A, CD 138, CD 1a, CD68, CD23, CD3, CD20, CD30, CD34, CD31, ALK1, desmin, and synaptophysin. Ki67 index was 60%–70% [Figures 2 and 3]. Above histopathological and IHC features are suggestive of cutaneous ES. In view of the above findings, the patient was planned for six cycles of palliative chemotherapy (vincristine, doxorubicin [Adriamycin®], and cyclophosphamide [VAC] regimen). The patient was also advised for daily dressing with locally applied antibiotic coverage. After three cycles of chemotherapy, a follow-up clinicoradiological assessment revealed a clinically significant reduction in the size of lesion. Radiological imaging (CECT brain, neck, thorax, and abdomen) after three cycles of chemotherapy revealed a 25%–50% reduction in the size of the lesion in left fronto–parieto–temporal region, measuring 6.1 cm × 2.7 cm × 6.4 cm posttreatment. Pleural-based soft-tissue lesion in the left anterior chest wall appeared mildly reduced in size (<25% reduction in size) [Figure 4]. The patient was advised to continue three more cycles of chemotherapy.

Discussion

Extraskeletal ES family of tumors (EESFT) most frequently occur in the deep soft tissues of children and young adults, such as the paraspinal muscles, chest wall, and the lower extremities. Sometimes, it can present in a superficial location either as a primary tumor or a metastasis from osseous or deep-seated EESFT. They are exceedingly rare and are limited to the skin and generally present as a single small lesion, circumscribed mid-to-deep dermis, or involving superficial subcutis.[1-5,7] As PCES is common in women, which is in accordance with our case, although the age group is higher and in a mixed-race patient (more common in white females). Normally, cutaneous tumors are considered of better prognosis when compared to the soft-tissue tumors, especially due to the possibility of early diagnosis, which is also the

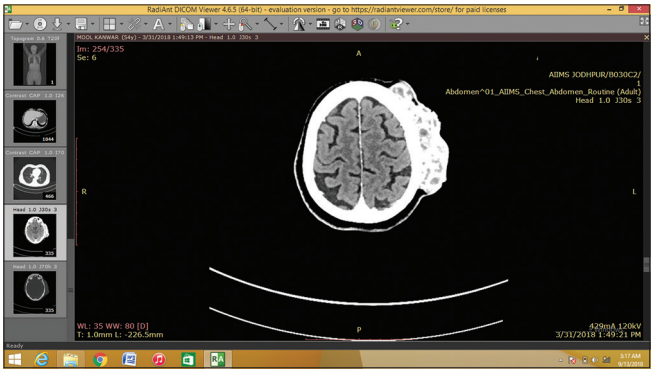

Figure 1: Prechemotherapy radiological image

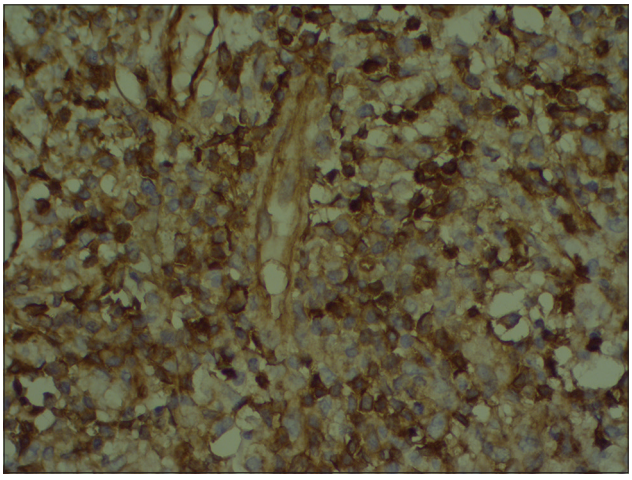

Figure 2: Immunohistochemistry CD-99- Positive

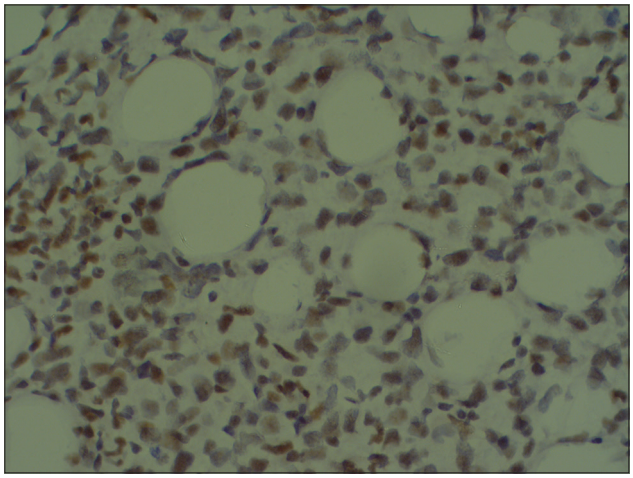

Figure 3: Immunohistochemistry FLI-1- Positive

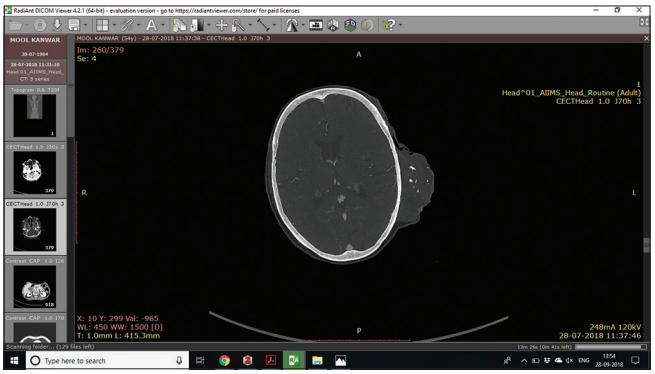

Figure 4: Postchemotherapy radiological image

scenario in our case. Cutaneous ES is often clinically and pathologically underdiagnosed as it is rare and morphologically similar to other cutaneous tumors.[1,4,5,8] The presence of metastases is very rare. In the reviewed reports, the majority of the patients did not present evidence of metastatic disease during minimum 16-year follow-up.[7] Clinically, morphologically, and genetically, extraskeletal ESs and PNETs share a lot of features, supporting the hypothesis that these two neoplasms are histogenetically related and they are widely considered as part of the same family of tumors.[9] Different diagnostic tools are aspiration cytology, histochemical stains, IHC, electron microscopy, cytogenetics, and molecular genetics of translocations. The ES is composed of small round cells expressing CD99 and molecular studies such as fluorescence *in situ* hybridization or real-time polymerase chain reaction shows a specific chromosomal translocation t (11;22) involving gene EWSR1 in chromosome 22q12 or a fusion or combination between EWSR1 gene and gene of ETS family.[6,7] PCES has better prognosis and behaves less aggressively than primary bone or soft-tissue ES with a survival rate of 91% in 10 years, which may be due to superficial location, small size, and easy access. One case of metastasis and one with local node involvement have been reported. The presence of metastasis is rare. The principles of treatment are currently similar to ES of the bone, including surgery with or without chemotherapy and/or radiotherapy, depending on its size and location.[4] Chemotherapy for these patients is based on the VAC alternating with ifosfamide and etoposide regimen.[10] Regimens used in subsequent lines include cyclophosphamide/topotecan, irinotecan/temozolomide, and docetaxel/gemcitabine.[6] Since our patient was already having metastasis, so palliative chemotherapy was planned. In conclusion, the current case reports presentation and treatment of PCES of scalp with lung metastases. Early awareness and wide resection followed by chemotherapy and radiotherapy might improve the long-term survival of patients with extraskeletal ES.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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