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Primary Cutaneous Ewing Sarcoma of the Scalp: A Case Report

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1. Introduction

Primary cutaneous extraskeletal Ewing Sarcomas (EWS) are extremely rare tumours limited to the skin [1–5]. Although they are less aggressive tumours than their counterparts, the intensity of chemotherapy given is the same [5]. When considering behavior

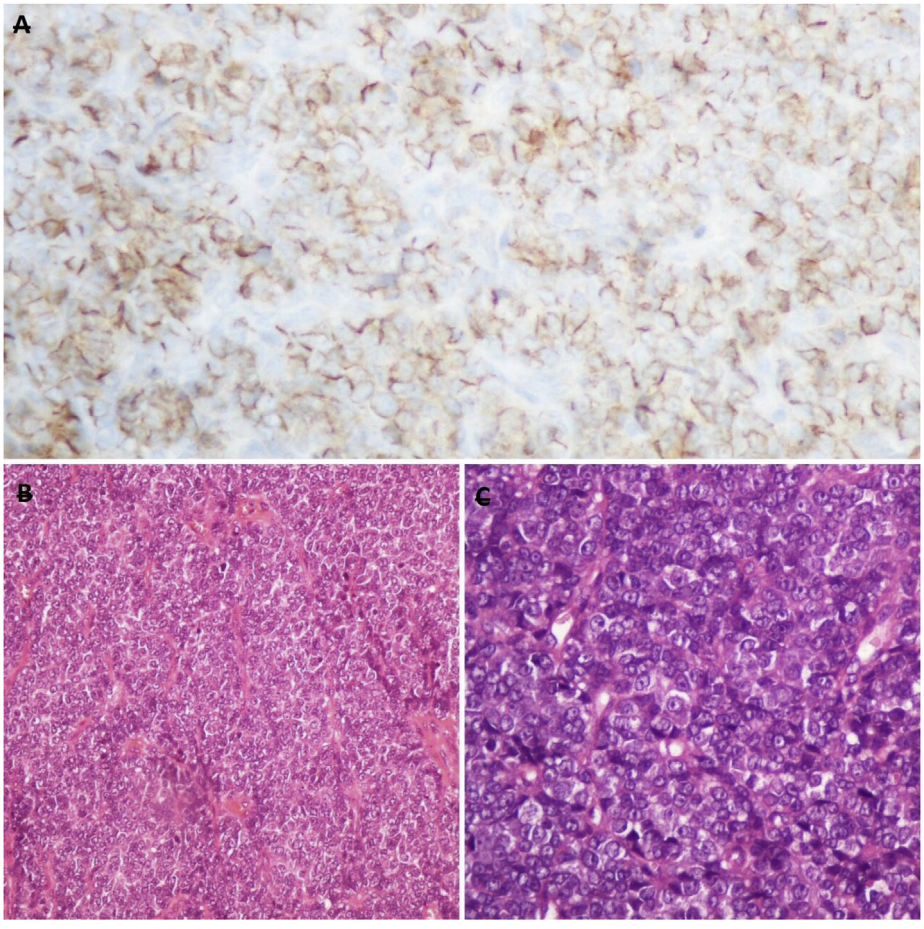

Fig. 1. A. Diffuse, membranous CD99 (Mic-2) positivity in tumoral cells, 400× magnification. B. Hematoxylin and eosin (H&E) stain, round cell tumor infiltration, 200× magnification.C. Hematoxylin and eosin (H&E) stain, round cell tumor infiltration, 400× magnification.

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of this tumor and chemotherapy related toxicity, intensity and duration of the therapy is controversial. Here, we present a boy diagnosed with primary cutaneous EWS and was treated by a short course of chemotherapy with no relapse until today.

2. Case presentation

An eight-year-old boy presented with a painless lesion on his scalp. On physical examination, a 4 × 4 cm size painless, mobile lesion was noted on the occipital region. Complete blood count and biochemical analysis were completely normal. Total excisional biopsy was done. The pathology specimen was limited to the dermis and immunohistochemical study showed diffuse membranous positive staining of CD99 (Fig. 1). The molecular analysis showed t (11; 22) involving gene *EWRS1*. These findings were consistent with EWS. Chemotherapy was started according to the Euro-Ewing (EE) 99 protocol. Hemorrhagic cystitis was observed in each cycle of chemotherapy. After the induction phase of 6 cycles of VIDE (vincristine, ifosfamide, doxorubicin, etoposide) remission was achieved. Radiotherapy was considered because of the presence of positive surgical margin, but patient's family refused the radiotherapy. Family also quit the remaining chemotherapy. There was no evidence of recurrence at 36th month of follow up.

3. Discussion

Cutaneous EWS is a rarely encountered tumor and only case series has been reported in literature. In the EE registry there were 24 (2.7%) cutaneous EWS cases reported between 1999 and 2012 [4]. Diagnosis is based on histological, immunohistochemical examination ($CD99^{+}$) and/or cytogenetic analysis, t (11; 22) [4]. The main differential diagnoses are melanoma, merkel cell carcinoma, cutaneous lymphomas, clear cell sarcoma, rhabdomyosarcoma and malignant rhabdoid tumor [2,3]. Prognosis of cutaneous EWS is better than its deep counterpart, with a 10-year probability of survival rate of 91% [1–5]. Giannatale et al. reported 5-year OS and EFS rates 93.8% and 88.5% respectively [4]. In this report 2 patients (2/56) (3.4%) had metastasis at diagnosis and died from progressive disease despite the chemotherapy. This report suggested that metastatic disease at diagnosis is clearly associated with a worse prognosis. Collier et al. collected 78 cutaneous EWS and it was shown that complete resection with negative margins significantly improved survival ($P = 0.037$) [5]. The excellent prognosis of localized cutaneous EWS raises the question of the optimal treatment for these patients. In Giannatale's study all patients but one received chemotherapy. Thirty of them received EE99 protocol, remaining 25 patiens received similar chemotherapy regimens. In the induction treatment, acute toxicities had been observed in all 30 patients, including hematological toxicity, infection, digestive, cardiac, and neurological toxicities. Chemotherapy doses were reduced in 57% patients due to myelosuppression. Twenty-seven of 56 patients (48%) received radiotherapy [5]. The effectiveness of radiotherapy as a local treatment in localized EWS is well known, but its role in the treatment of cutaneous EWS is still unclear [4]. Our patient completed induction of chemotherapy and didn't receive radiotherapy.

4. Conclusion

In conclusion further studies are warranted to determine whether a shorter and less intensive chemotherapy regimen could be safely used in this rarely encountered entity, with the aim of reducing chemotherapy-related toxicity.

Declaration of competing interests

The authors report no conflict of interest or funding to disclose.

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