SKIN

BRIEF ARTICLE

A Violaceous Mass of the Chest: An Unsuspected Diagnosis of Primary Ewing Sarcoma

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ABSTRACT

Ewing Sarcoma is the second most common primary bone cancer. To date, only a few cases of primary cutaneous Ewing Sarcoma have been published. We present a unique case of primary cutaneous Ewing Sarcoma, which presented as a blue to violaceous mass on a young woman's chest. The lesion clinically did not align with previous reports of primary cutaneous Ewing Sarcoma but pathology and immunohistochemical stains confirmed the diagnosis. Because primary cutaneous Ewing Sarcoma may masquerade as benign tumors, dermatologists must be suspicious of otherwise benign looking tumors.

CASE REPORT

A 30-year-old female with no past medical history presented to our dermatology clinic approximately three weeks after she noticed a rapidly growing asymptomatic plaque on her chest. On exam the patient was noted to have an indurated round non-mobile subcutaneous plaque with a light blue to violaceous color which measured 4.2 x 4 cm. No lymphadenopathy was found, and the exam was otherwise normal. The patient was biopsied the day of presentation and was prescribed doxycycline daily for 1 week. Pathologic examination of the biopsy specimen identified a high grade malignant small round blue cell tumor. Immunohistochemistry was performed and the tumor cells were found to be FLI-1, NKX2.2, CD99, Ki-67 and vimentin positive. Molecular studies were also performed and were positive for EWSR1 gene rearrangement. The diagnosis of primary cutaneous Ewing sarcoma was confirmed, and the patient was referred to oncology and general surgery. On follow up approximately one month after initial presentation, the tumor was re-examined and was found to have grown to 4.5 x 4.1 cm. The patient has since been started on chemotherapy with plans to have the tumor resected after her first round of chemotherapy is complete.

DISCUSSION

Epidemiology

With a peak incidence between 10 to 15 years of age, Ewing sarcoma (ES) is the second most common primary bone cancer. Almost a third of cases occur in children under 10, but the incidence in the elderly is unknown. The disease disproportionately affects males and has a predilection for

Caucasian patients.[1] On the other hand, primary cutaneous ES occurs more often in females and at later ages. It also tends to have a more favorable prognosis.[2]

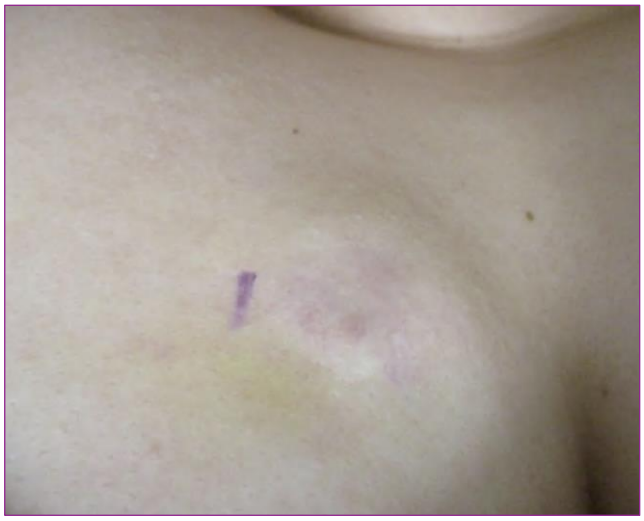

Figure 1. Indurated, non-mobile, violaceous plaque on the chest.

Presentation

Primary cutaneous Ewing sarcoma is only described in few case reports and series. Its clinical appearance can vary widely, making diagnosis difficult and necessitating biopsy, cytology, and immunohistochemistry. The primary lesion is composed of a mass in the dermis but is generally described as having pink or brown nodules with keratotic surfaces.[2] Our case presented with a mass in the dermis but with a blue to violaceous color being most apparent on the skin surface as can be seen in image 1.

Histopathology & Immunohistochemistry

Histopathologic evaluation of ES reveals small round cells with abundant cytoplasm and small nuclei. This characteristic is part of the family of tumors of childhood consisting of small blue round cell tumors. Periodic-acid-Schiff staining can also be useful to detect this cytoplasm due to its glycogen content. CD99 is an immunohistochemical biomarker highly sensitive for ES, but with low specificity. There are a host of other markers that may aid in identification (i.e. S-100, synaptophysin, CD45, vimentin, etc.), but their low specificity necessitates fluorescence in situ hybridization (FISH) or reverse transcription-polymerase chain reaction for diagnosis.[3] t(11;22)(q24;q12) is the most common gene rearrangement. EWS-FLI1 is a hybrid gene created in over 80% of cases of ES by the combination of the EWS and FLI1 genes on 22q12 and 11q24, respectively.[4]

Prognosis and Treatment

Treatment, regardless of the presence of metastasis, includes systemic chemotherapy and surgery or radiotherapy. The Euro Ewing 2008 treatment protocol has been used in cutaneous ES patients who can be appropriately treated with neoadjuvant chemotherapy. It consists of induction with 6 cycles of vincristine, ifosfamide, doxorubicin and etoposide (VIDE), followed by the appropriate prophylaxis.[6] Because smaller tumors may be seen more easily on the skin as compared to bone, primary cutaneous ES may be caught sooner. This potential for earlier detection has led to the idea of treatment regimens with less toxic medications, however clinical trials are still needed to confirm. Even given the earlier detection, roughly a tenth of these tumors already have metastasized by the time of presentation. For now, cutaneous ES is treated as aggressively as ES of the bone. Nonetheless, survival is far superior in cutaneous ES, with a 91% survival at 10 years.[7,] The risk of metastasis if undetected and consequently poorer prognosis makes early diagnosis and suspicion of a newly acquired lesion important for the clinician.

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