SKIN CANCER (OTHER THAN MELANOMA)

PRIMARY CUTANEOUS EWING SARCOMA

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Background: Ewing Sarcoma is a malignant small round cell tumor (neuroectodermal tumor). It is usually a primary bone tumor but its primary cutaneous site is extremely rare. It is mainly seen in children and young adults, though it sometimes affects elderly individuals. We report a case of primary cutaneous Ewing's sarcoma in a 53-year-old woman.

Observation: A 53-year-old woman presented with a bleeding lesion on his right heel, rapidly enlarging over the past 2 months. No other sites were affected and there were no associated systemic symptoms.
Physical examination revealed a solitary well-demarcated 1,5 cm exophytic firm pink keratotic and hemorrhagic lesion on her right heel.
Skin biopsy revealed a proliferation of small basophil round cells in the dermis and hypodermis. Immunostaining markers were positive for CD99, vimentin, and negative for epithelial (AE1/AE3, CK20), mesenchymal (MyoD, desmin), S100, melan A, HMB45, chromogranin A, synaptophysin, VIIIFactor, CD31, CD34, D2-40 markers. The fluorescence in situ hybridization analysis showed EWSR1 translocation (22q12). No visceral or bone injury were detected on imaging exams, and histological, immunohistochemical and molecular evaluation confirmed the diagnosis of primary cutaneous Ewing Sarcoma. The patient underwent wide surgical tumor resection and polychemotherapy and after 5 years follow-up no recurrences were detected.

Key Message: Primary cutaneous Ewing Sarcoma should be considered in the differential diagnosis of cutaneous 'small blue cell tumors'. Due to lack of specificity in its clinical presentation, histology and immunoprofile, diagnosis of primary cutaneous Ewing Sarcoma is difficult and many differential diagnoses must be considered. Detection of EWSR1 translocation (RT-PCR, FISH) can be useful for confirming the diagnosis of the neoplasm. Most cases are treated with wide surgical excision and adjuvant chemotherapy, and its prognosis seems better than bone Ewing Sarcoma.