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Clinica-Medical Image

Stewart Treves Syndrome: A Life Threatening Complication of Chronic Lymphedema

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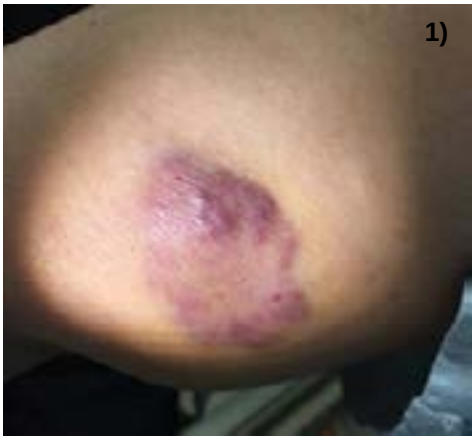

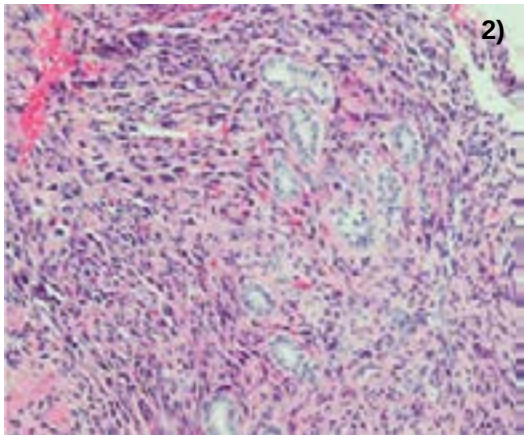

Figure 1: The figure shows the Right upper extremity (RUE) Papule.
Figure 2: Biopsy showed dermal proliferation of pleomorphic neoplastic cells.

Keywords:

Cancer; Lymph node; Radiation; Pain

Clinical Image

A 56-year woman with history of right breast cancer, initially treated with radical mastectomy with lymph node dissection, chemotherapy and radiation, presented with a six-month history of right upper extremity (RUE) swelling and pain. She had persistent RUE lymphedema since her cancer treatment. She developed a five-centimeter purple papule on her arm roughly three months prior to presentation (Figure 1). Biopsy showed dermal proliferation of pleomorphic neoplastic cells with round to irregular nuclear contours, inconspicuous to multiple prominent nucleoli and variable amphophilic cytoplasm, consistent with angiosarcoma (Figure 2). She underwent wide local excision of the mass and planned to receive adjuvant chemotherapy and radiation. Stewart Treves syndrome, first reported in 1948, refers to the association between chronic lymphedema and angiosarcoma, classically seen in women with breast cancer who develop lymphedema as a result of lymph node dissection during mastectomy. Approximately 400 cases of Stewart-Treves syndrome have been reported and the majority of them occurred in the ipsilateral upper extremities of breast cancer patients who have undergone axillary lymph node dissection. The pathophysiology is unclear, it is postulated that lymph stasis leads to the accumulation of proteins and induces local immunosuppression as well as vascular oncogenesis. Treatment options for Stewart-Treves Syndrome are limited. In early-stage angiosarcoma, wide excision is performed with the goal of obtaining negative surgical margins. Chemotherapy, immunotherapy and radiation therapy can be used as stand-alone treatment or as an adjuvant to surgery. Overall prognosis is poor, with a high rate of local recurrence and metastasis.

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