MONOPHASIC BREAST SYNOVIAL SARCOMA – CASE REPORT

Juliana Lopes de Aguiar Araujo¹, Ubiratan Wagner de Sousa¹, Fernanda Mabel Batista de Aquino¹, Luisa Gurgel de Lira², Iris Maria Rodrigues de Andrade Almeida³

¹Liga Norte Rio-Grandense Contra O Câncer – Natal (RN), Brazil.
²Universidade Potiguar – Natal (RN), Brazil.
³Universidade Federal do Rio Grande do Norte – Natal (RN), Brazil.

Introduction: Breast synovial sarcoma is extremely rare, with few cases described. It corresponds to 6–9% of soft tissue sarcomas and is more frequent in extremities (80%), trunk (8%), and abdomen (7%) in young adults. It usually does not affect the breast.

Objectives: To report a rare case of monophasic breast synovial sarcoma and provide data for the global literature.

Method/Case report: G.S.B., 97 years old, presented a 7 cm nodule in the left breast and negative axillary nodes. Ultrasound (US) revealed a heterogeneous nodule of 6.0 x 5.5 cm, BI-RADS 5. She did not have mammography. Core biopsy showed spindle cell neoplasm. Immunohistochemistry (IHC) indicated mesenchymal lesion, without differentiating stromal components of fibroepithelial tumor from the mesenchymal lesion. Rapid growth with ulceration and tumor bleeding were identified. Urgent mastectomy showed a malignant neoplasm of spindle cell pattern and high grade, with 12 cm, involved lateral margin, and 19 negative axillary lymph nodes. IHC of the surgical specimen indicated monophasic synovial sarcoma. Before the wide excision, she had a rapidly progressive recurrence in the sternum, making it non-resectable. During radiotherapy (RT), local progression was identified. She has been receiving chemotherapy (CT) with ifosfamide and Adriamycin. No evidence of distant disease was found after 9 months of diagnosis.

Results/Discussion: Synovial sarcoma corresponds to approximately 0.06% of all breast neoplasms, originating from their mesenchymal tissue, with variable epithelial differentiation. The term synovial sarcoma is inadequate, deriving from its frequent juxta-articular location. Its incidence is approximately 1.5 per 1 million individuals, with a mean age of 32 years and a male:female ratio of 1.2:1. The main histological subtypes are: classic biphasic and monophasic. Translocation t(X;18) (p11.2; q11.2) and expression of SYT/SSX gene fusion are present in more than 95% of cases. IHC shows an intense expression of vimentin and CD99, and focal of Bcl2, EMA, CKAE1-AE3, actin, and desmin, as well as negativity for S100, cytokeratins, hormone receptors, myosin, and caldesmon. The differential diagnosis is made with other spindle cell entities, such as fibromatosis, solitary fibrous tumor, myofibroblastic tumor, metaplastic carcinoma, and other sarcomas. Synovial sarcoma has a moderate response to chemotherapy with anthracyclines. The treatment includes wide surgical resection and RT. Metastases occur in about 50% of cases and are present at diagnosis in 16% to 25%; they are more frequent in the lung (75%), regional lymph nodes (15%), and bones (10%), tending to late recurrence and metastases. The 5-year disease-free survival is 60%.

Conclusion: The heterogeneity of the disease and its low incidence hinder prospective studies addressing therapeutic options with better long-term results.