HOW TO TREAT HISTIOCYTIC SARCOMA, A RARE MALIGNANT TUMOUR?

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Uvod: Histiocytic sarcoma (HS) is an extremely rare malignant neoplasm, accounting for less than 1% of all hemato-lymphoid neoplasms. The tumour cells are derived from monocyte/macrophage lineage and express histiocytic markers, including CD68, CD163, and Lysozyme.
Case report: A 60-year-old man presented with a right-sided neck tumour mass measuring 49mm×39mmx39mm. Microscopy of the lesion revealed a markedly pleomorphic tumour, composed of large cells with vesicular nuclei. Immunohistochemical staining of the tumour cells revealed that LCA S100, CD33, CD68, CD163, CD31, CD4, CD10 and Vimentin were positive.
Radiology evaluation pointed to enlarged nodes at axillae and inguinal regions. Bone marrow biopsy confirmed bone involvement. The patient received systemic chemotherapy (cyclophosphamide, doxorubicin, vincristine, etoposide and prednisone [CHOEP] regimen). After three cycles of chemotherapy, PET CT imaging revealed a hypermetabolic cervical lymph node (21× 24mm), SUV max 21.74. The chemotherapy regimen was changed to ESHAP (etoposide, methylprednisolone, cytarabine, cisplatin). Three cycles of therapy were applied and then an autologous stem cell transplant was performed. Control PET CT revealed hypermetabolic cervical lymph node (10x6x7 mm), SUV max 4,09. Radiotherapy had been applied on the right side of the neck, including radiation at the base of the tumour Control PET CT confirmed remission which persist for tree years of follow-up.
Conclusion: HS is an extremely rare malignant neoplasm of the monocytic/macrophage lineage, with no standardized chemotherapy regimen for the multisystemic disease. Metastatic patients have a more aggressive clinical course. High-dose chemotherapy including autologous stem cell transplantation may be an adequate therapy approach.

Key words: Histiocytic sarcoma, markers, CHOEP, ESHAP, stem cell transplantation