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15-year experience on vascular sarcomas: a retrospective study from a single institution

M.-C. Riesco-Martinez[1], L. Parrilla-Rubio[1], M.C. Munoz-Sanchez[1], A.-B. Enguita-Valls[2], R. Paz Ares[2], J.-A. Lopez-Martin[1]

[1] *Medical Oncology Department, Hospital 12 de Octubre, Madrid, Spain*

[2] *Pathology Department, Hospital 12 de Octubre, Madrid, Spain*

Introduction: Vascular sarcomas (VS) represent a rare group of soft-tissue border-line or malignant tumors that arise from blood vessels. Because of their low incidence, data on their treatment and prognosis are scarce. We aimed to evaluate the features, management and outcome patterns of this rare entity at a tertiary medical centre.

Methods: A retrospective cohort study was performed. All the files from patients with histological confirmation of intermediate or malignant VS, according to the WHO classification, treated at our institution from January 1999 to December 2014 were reviewed. Kaposi sarcoma pts were analyzed separately due to their distinctive etiopathogenesis. Demographic and histological characteristics were collected. Data on management, disease free survival (DFS) and overall survival (OS) were analyzed, and histological subtypes were compared.

Results: Data from 30 pts were collected. Median age at diagnosis was 63 years, (range 26-90). 50% were men (15/30). Histologic subtypes included: angiosarcoma (80%) and hemangioendothelioma (20%). The most common locations in both subtypes were: liver, lower limb and trunk with 27%(8/30), 23%(7/30) and 10%(3/30) pts respectively. Metastatic disease was present in 6/30 (20%) pts at diagnosis; 50%(3/6) angiosarcoma and 50% (3/6) hemangioendothelioma. Vimentin, CD31 and CD34 were the most common immunohistochemical markers, expressed in 81%, 86% and 50% of samples respectively. Main treatment was surgery, performed in 96% (23/24) of non-metastatic pts. R0 resection was achieved in 74% (17/23) of cases. Adjuvant radiotherapy and chemotherapy were administered in 29%(7/24) and 8%(2/24) of pts respectively. Only 2/30 (6.7%) pts received neoadjuvant treatment. Recurrence occurred in 37.5% (9/24) pts and was local in 55% of them. 70% of non-resectable pts received at least one line of chemotherapy. Bevacizumab alone or in combination with taxanes was the most frequent drug used in 57% (4/7) pts. Median OS was 6.5 years (95%CI 3.1- 9.9) for hemangioendothelioma and 3.5 years (95%CI 2.5-5.1) for angiosarcoma.

Conclusions: Surgery was the main treatment in VS. Despite high rates of R0 surgery recurrence was frequent. Therefore, additional treatment may be needed. Further studies on the role of adjuvant therapy are warranted.