

ABSTRACT # 131

CLINICAL PRESENTATION, MANAGEMENT AND PROGNOSIS OF INTRAVASCULAR LYMPHOMATOSIS (IVL): AN ONGOING CLINICO-PATHOLOGIC STUDY OF THE I.E.L.S.G. (IELSG #17 STUDY)

A.J.M. Ferreri, J.F. Seymour, S. Grisanti, G. Rossi, E. Berti, M.A. Pavlovsky, M. Martelli, S. Govi, M. Ungari, S. Ascani, S. Pileri, E. Pedrinis, C. Patriarca, G. Arrigoni, F. Facchetti, F. Cavalli, and M. Ponzoni on behalf of the International Extranodal Lymphoma Study Group (I.E.L.S.G.)

Background: IVL or angiotropic lymphoma is a rare entity characterized by exclusive or predominant growth of neoplastic cells within blood vessel lumina. Studies encompassing a relatively large group of IVL analyzed under therapeutic and biologic standpoint are lacking.

Purpose: Clinico-pathological characteristics regarding IVL of an ongoing retrospective I.E.L.S.G. series were reviewed to better understand clinical presentation, histopathologic features, adhesion and dissemination properties, course, and appropriate therapeutic approach.

Methods: 17 HIV-ve pts with *in vivo* or *post-mortem* diagnosis of IVL were reviewed.

Results: Median age was 73 ys (51-90); M:F ratio 1.4; ECOG-PS>1= 11. Three pts had a previous or concomitant malignancy (gastric MALT lymphoma in one). Neurologic symptoms, fever and cutaneous lesions were the most common presentation. B-symptoms were observed in 10 cases (fever of unknown origin in 9). *Post-mortem* diagnosis was made in 2 cases. *In vivo* diagnosis were performed incidentally during surgical resection of other tumors in 2 cases, on bone marrow biopsy in 2, or by biopsy of lesions from skin (n=5), brain (n=2), uterus (n=2), gallbladder (n=1), and kidney (n=1). Stage of disease (Ann Arbor) at diagnosis was IE in 8 cases and IVE in 9 (clinical staging in one, bone marrow involvement in 2, multiple cutaneous lesions in 3, *post-mortem* staging in 3). Anemia was observed in 14 cases, leukopenia in 4, thrombocytopenia in 3, elevated ESR in 11, elevated LDH serum level in 10, monoclonal component in 3, and hypoalbuminaemia in 5.

Ten pts were treated with CHOP or CHOP-like chemotherapy (CHT) (5 with stage-I disease; followed by high-dose CHT + PBSCT in one), one pt with cutaneous disease was treated with steroids alone, and 4 pts did not receive any treatment due to early death or refusal. Among pts treated with CHT, two died of toxicity, 4 had progressive disease, two experienced relapse after initial remission, and the other two pts (both with stage IE) are alive and relapse-free at 47 and 68 months from diagnosis. Among pts who did not receive CHT, 4 died of renal or lung complications at 1 - 24 months, and one was lost to follow-up after one month. Failures involved mainly primary sites of disease; the median time to treatment failure was 4 months, with a one-yr failure-free survival of 24±11%. Two of the 4 pts who received salvage therapy achieved a second response and are alive with disease at 9 and 21 months. The pt treated with high-dose CHT is alive and NED at 47 months from diagnosis. Five pts are alive (2 NED) with a median follow-up of 21 months (one-yr OS: 42±13%). Pts with cutaneous disease exhibited a better outcome than pts with CNS involvement (one-yr OS: 66±15% vs. 14±13%; p=0.06).

Conclusions: Preliminary results suggest that IVL is an aggressive and usually disseminated disease that affect elderly pts, with a slight male predominance, and presenting poor PS, B-status, anemia, elevated ESR, and high LDH serum level. Diagnosis is often incidental. Although presentation is consistent with multiorgan involvement, staging work-up is often unable to detect systemic disease. Brain and skin are the most frequently involved organs, while lymph nodes are usually spared. Even though aggressive CHT may produce prolonged remission in some cases, survival is disappointing. A larger number of pts is needed to better define biological characteristics, clinical course and prognostic factors as well as optimal therapeutic management.

Mail to: <andres.ferreri@hsr.it> or <maurilio.ponzoni@hsr.it>

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