

CLINICAL FEATURES OF THE WESTERN AND ASIAN FORMS OF INTRA-VASCULAR LYMPHOMA (IVL) VARIES ACCORDING TO THE PRESENCE OF HEMOPHAGOCYTIC SYNDROME (HPS) AND NOT TO THE GEOGRAPHICAL AREA (IELSG 17)

A.J.M. Ferreri, G.P. Dognini, E. Campo, E. Zucca, M. Martelli, A. De Renzo, C. Doglioni, M. Ferracci, C. Montalban, A. Tedeschi, A. Pavlovsky, A. Morgan, L. Uziel, S. Ascani, U. Gianelli, C. Patriarca, F. Facchetti, L. Mazzucchelli, F. Cavalli, M. Ponzoni (Milan, Italy)

Background Published data suggest the existence of some clinical differences between IVL patients diagnosed in Asian and Western countries.

Aim: To explore potentially different clinical forms of IVL by comparing the clinical features of the largest cumulative series of IVL patients diagnosed in Western countries and three subgroups of IVL patients diagnosed in different Asian countries and published in the English literature.

Methods Clinical records and pathological material of 45 HIV-negative patients with IVL diagnosed in 8 Western Countries (Western-IVL) were reviewed. Clinical features of this series were compared with 282 previously reported cases of IVL diagnosed in Western countries (Western-IVL) and with 120 previously reported cases of IVL diagnosed in Japan (n=86) and other Asian Countries (n=34). Analysis was performed according to the presence of HPS.

Results HPS was absent in our patients; it was diagnosed in 5 (2%) previously reported cases of Western-IVL (p= 0.37), in 38 (44%) Japanese patients (p= 0.00001) and in 4 cases (12%) diagnosed in other Asian countries (p= 0.03).

Analysis of differences in clinical presentation and laboratory findings included four subgroups: our 45 Western-IVL patients, 38 Japanese patients with IVL and HPS (J-HPS), 48 Japanese patients with IVL without HPS (J-IVL) and 30 patients with IVL without HPS diagnosed in Asian countries other than Japan (Eastern-IVL). Median age was very similar among studied subgroups, oscillating between 62 and 69 years, with a constant slight prevalence among males. As reported in the table, there were no significant differences in presenting symptoms, sites of disease or laboratory findings among Western-IVL, J-IVL and Eastern-IVL patients. Conversely, stage-IV disease, fever of unknown origin, involvement of liver, spleen, bone marrow or lung, fatigue, jaundice, thrombocytopenia, increased serum levels of hepatic enzymes as well as a concomitant extravascular lymphoma were significantly more common among the 38 J-HPS patients in comparison with the other groups. Conversely, skin and central nervous system involvement was significantly more rare in J-HPS patients. No significant differences were observed in terms of anemia, leucopenia, monoclonal component, and peripheral blood involvement. In patients treated with anthracycline-based chemotherapy (21 from our Western-IVL series and 27 from J-HPS series), complete remission rate was 52% and 58% (p= 0.92), with a 2-year overall survival of 45±11% and 22±8% (p= 0.04), respectively.

Conclusions The association between IVL and HPS is anecdotally diagnosed outside of Japan. IVL significantly varies in clinical features and laboratory findings according to the presence of HPS and not to the geographical area. Patients with IVL but without HPS diagnosed in Western countries, Japan and other Asian countries display similar characteristics and could be considered as forming part of a "classical form" of IVL. J-HPS patients display numerous clinical differences with respect to the classical form and could be considered as a "HPS-related variant" of IVL. When treated with anthracycline-based chemotherapy, both variants exhibits a worse prognosis, specially in HPS-related cases; thus, rendering advisable treatment intensification. An extensive phenotypic and molecular characterization is needed to confirm whether these clinical differences might reflect discordant biological entities within IVL.

	Western (n=45)	J-HPS (n=38)	p	J-IVL (n=48)	p	Eastern (n=30)	p
Fever	19(42%)	34(89%)	0.00001	20(42%)	0.96	18(60%)	0.13
Stage IV	34(76%)	37(97%)	0.004	45(94%)	0.19	25(83%)	0.42
Skin	17(38%)	1(3%)	0.0001	12(25%)	0.18	7(23%)	0.19
CNS	18(40%)	8(21%)	0.04	25(45%)	0.24	11(37%)	0.77
Liver	12(27%)	25(66%)	0.0004	15(31%)	0.63	13(43%)	0.13
Spleen	11(24%)	29(76%)	0.0001	10(21%)	0.68	10(33%)	0.40
Lymph n.	4(9%)	2(5%)	0.68	2(4%)	0.43	9(30%)	0.02
Lung	8(18%)	14(37%)	0.05	13(27%)	0.28	15(50%)	0.003
B. marrow	14(31%)	28(74%)	0.001	16(33%)	0.82	4(13%)	0.10
Thrombocyt	16(36%)	28(74%)	0.005	6/34(18%)	0.007	9/25(36%)	0.97
High LDH	29/34(85%)	36(95%)	0.17	31/33(94%)	0.43	21/21(100%)	0.14
High ALT	4(9%)	10(26%)	0.04	3(6%)	0.71	8(27%)	0.06
High bilirubin	2(4%)	11(29%)	0.004	0(0%)	0.23	1(3%)	1.00



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