Supplementary Table 1 Comparison of clinical characteristics of our cohort and other two cohorts.

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| Characteristics | At diagnosis, n (%) |  | cf*BRAF*V600E |  |  | lt*BRAF*V600E |
| our cohort | French cohort 1# | French cohort 2\* |  | our cohort | *P* | French cohort 1# | *P* |  | our cohort | *P* | French cohort 2\* | *P* |
|  | - (%) | + (%) | - (%) | + (%) |  | - (%) | + (%) | - (%) | + (%) |
| Total | 140 | 48 | 315 |  | 60.0 | 40.0 |  | 56.8 | 43.2 |  |  | 40.0 | 60.0 |  | 45.4 | 54.6 |  |
| Sex |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| Male | 76 (54.3) | 25 (52.1) | 167 (53.0) |  | 59.5 | 46.4 | 0.128 | 44.0 | 52.6 | 0.760 |  | 64.3 | 47.6 | 0.052 |  |  |  |
| Female | 64 (45.7) | 23 (47.9) | 148 (47.0) |  | 40.5 | 53.6 | 56.0 | 47.4 |  | 35.7 | 52.4 |  |  |
| Median age at diagnosis, years | 2.3 | 2.1 | 3.2 |  | 3.6 | 1.4 | <0.001 | 2.8 | 1.0 | 0.009 |  | 3.2 | 1.7 | 0.106 | 3.7 | 2.5 | 0.01 |
| Clinical classification |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| SS LCH | 64 (45.7) | 21 (43.8) | 196 (62.2) |  | 57.1 | 28.6 |  | 60.0 | 15.8 |  |  | 50.0 | 42.8 |  | 56.1 | 43.9 |  |
| MS RO－LCH | 49 (35.0) | 12 (25.0) | 70 (22.2) |  | 35.7 | 33.9 | <0.001 | 36.0 | 10.5 | < 0.001 |  | 42.9 | 29.8 | 0.010 | 38.6 | 61.4 | <0.001 |
| MS RO+ LCH | 27 (19.3) | 15 (31.2) | 49 (15.6) |  | 7.1 | 37.5 |  | 4.0 | 73.7 |  |  | 7.1 | 27.4 |  | 12.2 | 87.8 |  |
| Involvement |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |  |
| Bone | 123(87.8) | 38 (79.2) | 257 (81.6) |  | 90.5 | 83.9 | 0.245 | 84.0 | 73.7 | 0.470 |  | 91.1 | 85.7 | 0.342 |  |  |  |
| Skin | 39 (27.8) | 27 (56.3) | 113 (35.9) |  | 16.7 | 44.6 | <0.001 | 40.0 | 84.2 | 0.005 |  | 16.1 | 35.7 | 0.011 | 23.0 | 77.0 | <0.001 |
| Liver | 23 (16.4) | 12 (25.0) | 37 (11.7) |  | 4.8 | 33.9 | <0.001 | 4.0 | 57.9 | < 0.001 |  | 5.4 | 23.8 | 0.004 | 10.8 | 89.2 | <0.001 |
| Spleen | 15 (10.7) | 12 (25.0) | 36 (11.4) |  | 2.4 | 23.2 | <0.001 | 4.0 | 57.9 | < 0.001 |  | 0.0 | 17.9 | 0.001 | 11.1 | 88.9 | <0.001 |
| Hematological | 12 (8.6) | 14 (29.2) | 37 (11.7) |  | 2.4 | 17.9 | 0.003 | 0 | 73.7 | < 0.001 |  | 3.6 | 11.9 | 0.124 | 10.3 | 89.7 | 0.003 |
| Pituitary | 11 (7.9) | 5 (10.4) | 48 (15.2) |  | 8.3 | 7.1 | 1.000 | 12.0 | 10.5 | 1.000 |  | 5.4 | 9.5 | 0.526 | 27.1 | 72.9 | 0.007 |
| CNS | 8 (5.7) | 2 (4.2) | 22 (7.0) |  | 4.8 | 7.1 | 0.713 | 4.0 | 5.3 | 1.000 |  | 5.4 | 6.0 | 0.745 | 25.0 | 75.0 | 0.05 |
| Lung | 31 (22.1) | 2 (4.2) | 37 (11.7) |  | 17.9 | 28.6 | 0.135 | 0 | 5.3 | 0.430 |  | 23.3 | 21.4 | 0.803 |  |  |  |
| Lymph nodes | 16 (11.4) | 6 (12.5) | 27 (8.6) |  | 10.7 | 12.5 | 0.745 | 4.0 | 26.3 | 0.070 |  | 12.5 | 10.7 | 0.790 |  |  |  |
| Follow-up, years; median (range) | 2.6 (0.1-3.8) | 2.2 (0.4-11.7) | 3.2 (0.0-17.0) |  |  |  |  |  |  |  |  |  |  |  |  |  |  |

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\*Heritier S, Emile JF, Barkaoui MA, Thomas C, Fraitag S, Boudjemaa S, et al. BRAF Mutation Correlates With High-Risk Langerhans Cell Histiocytosis and Increased Resistance to First-Line Therapy. J Clin Oncol. 2016;34:3023-30.

-: negative; +: positive.