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Commentary

Castleman disease, TAFRO syndrome, idiopathic plasmacytic lymphadenopathy, and autoimmune disease

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Although Castleman disease (CD), first described by Castleman in 1954[1] and termed multicentric CD by Frizzera *et al*. in 1983,[2] is a famous disease, CD was regarded as being rare and complicated. Several subtypes (diseases) have been included in CD and finally, in 2017, the first international consensus diagnostic criteria for idiopathic (KSHV/HHV-8 negative) multicentric Castleman disease (iMCD) were published, and CD was considered a heterogeneous group consisting of unicentric CD (UCD), HHV8-associated MCD, iMCD-NOS, iMCD-TAFRO, and POEMS-associated MCD.[3] The concept of CD seems to be clear; however, we still have several questions regarding the relationships among CD and related diseases, especially in Japan.

Idiopathic plasmacytic lymphadenopathy with polyclonal hyper immunoglobulinemia (IPL) was first described by Mori *et al*. in 1980,[4] three years before Frizzera's paper on MCD was published. This disease was further described by Kojima *et al*. in the 2000s and he subdivided MCD into IPL and Non-IPL, the latter including autoimmune disease-associated lymphadenopathy.[5] On the other hand, TAFRO syndrome (thrombocytopenia, ascites/anasarca, fever, reticulin fibrosis, renal dysfunction, and organomegaly) was first described by Takai *et al*. in 2010.[6] Since its pathological relationship to CD was indicated by Kojima *et al*. in 2008,[5] what Kawabata *et al*. referred to as Castleman-Kojima disease,[7] is now termed iMCD-TAFRO. Moreover, TAFRO syndrome is clinically known to include patients with autoimmune diseases.

In this issue, we reviewed CD and related diseases for understanding the relationships among these diseases. Nishimura M.F. *et al*. and Nishimura Y. *et al*. review 'Historical and pathological overview of Castleman disease' and 'International definition of iMCD-TAFRO: future perspectives', respectively.[8,9] Takeuchi K. discusses IPL with an original paper by Mori translated to English.[10] Koga T. and Kawakami A. *et al*. describe the clinical features of iMCD-TAFRO.[11] Kawakami A. is proceeding with Japanese research groups for CD and TAFRO syndrome.[12,13] Finally, Nakazato Y *et al*. describe the pathology of autoimmune diseases.[14]

Last of all, I hope this special issue will be useful for the readers of JCEH and I would like to dedicate these discussions to the late professor Masaru Kojima, who was most familiar with non-neoplastic diseases of the lymph nodes and contributed with many writings.

REFERENCES

1 Castleman B, Towne VW. Case 40011. N Engl J Med. 1954; 250: 26-30.
2 Frizzera G, Banks PM, Massarelli G, Rosai J. A systemic lymphoproliferative disorder with morphologic features of Castleman's disease. Pathological findings in 15 patients. Am J Surg Pathol. 1983; 7: 211-231.
3 Fajgenbaum DC, Uldrick TS, Bagg A, *et al*. International, evidence-based consensus diagnostic criteria for HHV-8–negative/idiopathic multicentric Castleman disease. Blood. 2017; 129: 1646-1657.
4 Mori S, *et al*. J Jpn. Soc Res. 1980; 20(suppl): 55-65. [in Japanese]
5 Kojima M, Nakamura N, Tsukamoto N, *et al*. Clinical implications of idiopathic multicentric castleman disease among Japanese: a report of 28 cases. Int J Surg Pathol. 2008; 16: 391-398.
6 Takai K, Nikkuni K, Shibuya H, Hashidate H. [Thrombocytopenia with mild bone marrow fibrosis accompanied by fever, pleural effusion, ascites and hepatosplenomegaly]. Rinsho Ketsueki. 2010; 51: 320-325. [in Japanese with English abstract]
7 Kawabata H, Takai K, Kojima M, *et al*. Castleman-Kojima disease (TAFRO syndrome) : a novel systemic inflammatory disease characterized by a constellation of symptoms, namely, thrombocytopenia, ascites (anasarca), microcytic anemia, myelofibrosis, renal dysfunction, and organomegaly : a status report and summary of Fukushima (6 June, 2012) and Nagoya meetings (22 September, 2012). J Clin Exp Hematop. 2013; 53:

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57-61.
8 Nishimura, et al. J Clin Exp Hematop. 2022; 62.60-72
9 Nishimura, et al. J Clin Exp Hematop. 2022; 62.73-78
10 Takeuchi K. J Clin Exp Hematop. 2022; 62.79-84
11 Koga T, et al. J Clin Exp Hematop. 2022; 62.85-90
12 Research based on international consensus formation for the formulation and further improvement of clinical practice guidelines for Castleman's disease, TAFRO syndrome and related diseases.
13 Practical application research toward the establishment of precision medicine infrastructure of Castleman disease and TAFRO syndrome using integrated registries.
14 Nakazato, et al. J Clin Exp Hematop. 2022; 62. in preparation