Castleman Disease with the Experience over 17 Years

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INTRODUCTION

- Castleman disease (CD) encompasses several distinct clinicopathological disorders at the intersection of haematology, immunology, oncology, rheumatology and virology that share a spectrum of histopathological features.
- An international collaborative working group has reached consensus definitions and classification, defining diagnostic criteria for CD which enables to clinicians reaching the proper diagnose.

METHOD(S)

- All patients with a biopsy proven histopathological characteristics of CD diagnosed at Ege University Hospital between 2000-2017 years were reviewed for analysis.
- Clinical and laboratory datas were collected retrospectively.
- The patients were divided into two main groups based on the anatomical distribution of the disease: Unicentric CD (UCD) and multicentric CD (MCD). Also MCD were divided into two groups: HHV8 positive MCD and idiopathic MCD.

RESULT(S)

Table: Patient characteristics.

		UCD			MCD	
Table 1: CD. Patient characteristics	**Total**	**Single lesion**	**Single station**	**Total**	**HHV-8+MCD**	**iMCD**
N	16	12	4	14	7	7
Sex male/female	7/9	6/6	1/3	10/4	5/2	5/2
Age (median), years	41.8	37.3	55.5	55.4	54.6	52.6
Lymph node localisation						
Mediastinum	3	2	1	12	7	5
Abdominal	3	2	1	12	5	7
Cervical	6	6	-	13	7	6
Axillary	3	1	2	13	7	6
Inguinal	-	-	-	11	5	6
Extranodal	1	1	-	1	1	-
Delay to diagnosis (median, months)	1.8	1.25	3.75	7.2	11.2	3.1
Fever	1	-	1	11	7	4
Complications						
Splenomegaly	-	-	-	12	6	6
Oedema/effusion	-	-	-	4	3	1
Lung involvement	-	-	-	3	2	1
Skin lesions	1	-	1	10	6	4
Kidney involvement	1	-	1	5	2	3
Paraneoplastic pemfigus	-	-	-	-	-	-
Polyneuropathy	1	-	1	6	3	3
AIHA	-	-	-	1	1	-
AITP	-	-	-	1	1	-
Leucocyte count, $\times 10^9$ /L (min-max)	7760 (5650-11000)	7627 (5650-11000)	8162 (6170-9180)	9044 (3630-22800)	7960 (3630-16420)	10128 (5730-22800)
Heaemoglobin, g/dL (min-max)	12.6 (7.9-16.2)	13.4 (12-16.2)	8162 (6170-9180)	10.7 (5.6-15.2)	11 (7.7-15.2)	10.5 (5.6-14.3)
Platelet count, 10^9/L (min-max)	318 (78-655)	301 (220-461)	370 (78-655)	229 (56-564)	186 (70-277)	272 (56-564)
CRP, mg/dl (min-max)	1.7 (0.1-14)	0.3 (0.1-0.6)	5.9 (0.3-14)	7.4 (0.1-22)	6 (0.3-12)	8.7 (0.1-22)
Serum albumin, g/L (min-max)	4 (2.1-5.1)	4.1 (3.3-5.1)	3.4 (2.1-4)	3.1 (2-4.5)	2.9 (2-4.5)	3.3 (2.3-4.5)
Gammaglobulin, g/L (min-max)	3 (2-4.4)	2.9 (2-3.8)	3.5 (2.9-4.4)	4 (2.6-5.9)	4.1 (3-5.7)	3.8 (2.6-5.9)
Monoclonal gammopathy	-	-	-	4	2	2
LDH > normal	2	-	2	11	6	5
Ferritin > 5 times normal	1	-	1	10	6	4
DAT	-	-	-	4	2	2
Follow-up (months)	54	55.5	49.7	37.2	38.2	36.1
Deaths	2	-	2	3	2	1

CONCLUSION(S)

- CH is a very rare lymphoproliferative disease which should be kept in mind in the differential diagnosis with asymptomatic and localized lymphadenopathies or widespread lymphadenopathies with severe systemic symptoms.
- Future studies should be multicentred and collaborative in order to evaluate significant numbers of patients and to establish up to date and effective treatment protocols for this rare but potentially life-threatening disorder.

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