The clinical picture of Castleman disease: a systematic review and meta-analysis of almost 2000 patients

**Introduction**

Castleman disease (CD) encompasses a spectrum of rare disorders with characteristic histopathological features, including unicentric (UCD), idiopathic multicentric (iMCD), and cases associated with human herpes virus 8 (HHV8+ MCD). As treatment options for each type of CD vary, it is important to be able to distinguish their clinical presentation and determine the correct diagnosis.

To our knowledge, there are no major systematic reviews that describe and compare the clinical features of different CD subtypes. Whether these cases represent a distinct entity or are part of a spectrum of diseases is unresolved.

**Methods**

We performed a systematic review of publications reporting 35 cases of CD between 1995 and 2021. Publications were cross-checked by study location, data collection period, and investigators to identify and exclude duplicated publications.

For each subtype of CD within each study, we extracted data on demographics, clinical symptoms, and laboratory parameters, as stated in the international consensus diagnostic criteria for iMCD.

We estimated the mean percentage of patients meeting each diagnostic criterion for each CD subtype using meta-analyses conducted with random effects logistic regression models and exclude duplicated patients.

**Pediatric UCD patients had more signs of systemic inflammation compared with adult UCD patients**

Five publications with 88 pediatric UCD cases were included in the meta-analyses. Elevated C-reactive protein was reported in 45.1% of pediatric UCD patients (95% CI 22.7–70.5%, n=219) versus 38.2% (38.9–55.9%, n=715) in adult UCD patients.

**Pulmonary findings were observed in 15.3% of UCD cases**

Except for cutaneous findings, patients with UCD had significantly lower rates of symptoms vs. those with either iMCD or HHV8+ MCD.

**Conclusions**

We found several distinct differences between iMCD and HHV8+ MCD, which may reflect heterogeneity in underlying pathophysiology and/or different comorbidity burdens of these CD subtypes.

**References**


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