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Leukaemia Section

Mini Review

Castleman's disease

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Identity

Alias

Angiofollicular lymph node hyperplasia

Clinics and pathology

Phenotype/cell stem origin

The disease appears to be polyclonal in origin in the majority of cases; however evidence for clonal expansion was documented in some cases, possibly representing transformation into non Hodgkin's lymphoma. In approximately 1/3 of the cases studied a monoclonal IgH rearrangement was documented. A minor T-cell clone, mostly in a polyclonal background, was also documented in some cases.

Epidemiology

The disease is very rare.

Clinics

The disease may present as a solitary mass frequently occurring in the mediastinum or as a systemic disorder (multicentric Castleman's disease) with diffuse adenopathies, systemic symptoms and recurrent infections. Splenomegaly, hepatomegaly and neurologic symptoms may occur frequently as is the case with autoimmune manifestations.

Pathology

There are two variants: the hyaline-vascular and the plasma cell subtype. In the former subtype there are shrunken germinal centres with concentric expansion of the mantle zones with eosinophils and hyalinization around the vessels; in the latter subtype an extensive infiltrate by plasma cells is seen in the interfollicular areas. Some patients may be infected by human herpervirus-8 which may induce interleuchin-6 (IL6) overproduction. IL6 is believed to play an essential role in the pathogenesis of the disease.

The hyaline-vascular type is usually diagnosed in asymptomatic patients whereas systemic symptoms are present in the majority of patients with the plasma cell subtype. Patients with "multicentric" Castleman disease show histologic features consistent with the plasma cell subtype.

Treatment

The patients can be treated by surgical excision if the mass is localized. Steroid treatment is recommended in cases with disseminated disease and combination chemotherapy utilized for lymphoma must be reserved to unresponsive patients. Some patients with HIV associated Castleman's disease were successfully treated with the anti CD20 monoclonal antibody or with the antiviral agent ganciclovir targeting the HHV-8.

Prognosis

The prognosis varies greatly depending on the histologic type and disease presentation. If the disease is localized, surgery with or without radiotherapy may be curative. Those patients with multicentric disease who fail to respond to steroid treatment have a serious disease.

Cytogenetics

Cytogenetics morphological

Many of the cases so far studied showed a normal karyotype.

Occasional abnormalities were found in a few patients.

One case with the hyaline vascular type showed a t(1;6)(p11;p11), a del(7)(q21q22) and a del(8)(q12q22). In this patient no clonal expansion of lymphoid cells

was present, suggesting that the clonal proliferation involved dysplastic stromal cells.
Another patient was shown to carry a clonal abnormality in CD21-positive follicular dendritic cells. Abnormal chromosomes in this patient were add(1)(q21), der(6)t(6;12)(q23;q15), add(7)(p22), -9, inv(9)p11q13), del(12)(q15).
One patient carried a t(7;14)(p22;q22).

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