Castleman Disease in an Infant Patient: Case Report and Review of Literature

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Abstract

Background & Objectives: Castleman disease (angiofollicular lymphoid hyperplasia) is a rare lymphoproliferative disorder with a frequent mediastinal location, but may occur in any lymph node or extranodal site. It usually appears in young adults and rarely occurs in childhood and even rarer in infants and toddlers. There are only 100 pediatric cases described in the literature.

Methods: Online medical journal databases were searched for pediatric patients with Castleman disease. Results: This is the case of a 3 year old girl who presented a progressively growing right lateral-cervical mass since the age of 3 months.

In 2010 and 2012, two magnetic resonance imaging studies revealed an hemangiomma-like lesion which was refractory to medical therapy. In 2013 a mass at level III of the neck was resected and a neck dissection of levels II, III and IV was performed. The resulting histopathological diagnosis was Castleman disease of hyaline-vascular variant. Conclusion: Castleman disease has rarely been reported in infants. The cervical location, at whatever age, is far rarer than the mediastinal form. Most cases of cervical lymphadenopathy in children are not significant, but some are life threatening.

Castleman disease should be included in the differential diagnosis in persistent childhood lymphadenopathy.

Case Report

We describe the case of a 3 year old girl who presented a progressively growing right cervical mass since the age of 3 months. In 2010 and 2012 two magnetic resonance imaging studies revealed an hemangiomma-like lesion which was refractory to medical therapy. In 2013 a mass at level III of the neck was resected and a neck dissection of levels II, III and IV was performed. We received 19 lymph nodes from the right neck. The largest one measured 6 cm in greatest dimension. The cut surface was pink tan and firm. Histologically, lymphoid follicles were increased throughout the cortex and medulla, most containing two or more small germinal centers (so called “twinning”). Some of the follicles were surrounded by a broad mantle zone composed of concentric rings of small lymphocytes (so-called “onion skin”). Immunohistochecmistry was done and BCL6 highlighted the germinal centers, while CD79a demarcated the broad mantle zones.

Figure 1: Lymphoid follicles with multiple germinal centers. (H&E stain) (20X).

Figure 2: Sclerotic blood vessel penetrating a germinal center. (Lupin stain) (4X).

Figure 3: Lymphoid follicle with multiple germinal centers. (H&E stain) (40X).

Figure 4: Sclerotic blood vessel penetrating a germinal center. (Lupin stain) (10X).

Figure 5: Broad mantle zone demarcated by CD79a. Immunostain, (40X).

Figure 6: Germinal center highlighted by BCL6 immunostain. (40X).

The resulting histopathological diagnosis was Castleman disease of hyaline-vascular variant.

Discussion

Castleman disease, also known as angiofollicular lymph node hyperplasia or giant lymph node hyperplasia, was originally described as solitary lesions confined to the mediastinum, which is still the most frequent site of involvement. Castleman disease can occur anywhere throughout the lymphatic system. The most common sites include the mediastinum (60%), neck (14%), abdomen (11%), and axilla (4%). The disease is most commonly localized but rarely may be multicentric or systemic.

There are three forms: The hyaline vascular type, the plasma cell type and the systemic (multicentric) type. More than 90% of cases of Castleman disease have the hyaline vascular histologic appearance. The incidence of the disease is the same for male and female. Patients are generally asymptomatic unless the tumor mass partially obstructs a vital structure. Laboratory abnormalities, other than elevated LDH levels in a subset of patients, are rare. In these cases the lesions frequently occur as a single mass and range in size from 1.5 cm to 16 cm. The plasma cell variant accounts for 10% to 20% of the localized cases. These patients tend to be older than those with the hyaline vascular variant. The mediastinum tends to be involved less frequently than in patients with the hyaline vascular variant. Patients with the plasma cell variant can present with anemia, thrombocytopenia, and elevated serum interleukin 6 (IL6) levels. The multicentric variant is associated with human immunodeficiency virus (HIV) which is linked to human herpes virus 8 (HHV8) and Kaposi sarcoma.

B type symptoms (fever, night sweat, weight loss) occur in over 95% of these patients. These patients can also present splenomegaly, hepatomegaly, and multiple laboratory abnormalities (polycunal hypergammaglobulinemia, anemia, elevated ILA, thrombocytopenia, etc.). Furthermore, the multicentric variant can be associated with POEMS syndrome (peripheral neuropathy, organomegaly, endocrinopathy, monoclonal M protein, and skin lesions) and may predispose the patient to develop malignant lymphoma.

In children, Castleman disease has a more benign prognosis. It also has a different propensity for certain anatomic sites compared to adults, most commonly affecting the chest (33%), abdomen (30%), neck (23%), and axilla (7%). Most cases of cervical lymphadenopathy in children are not significant, but some are life threatening. Castleman disease should be included in the differential diagnosis in persistent childhood lymphadenopathy.

References