Cervical Posterior Triangle Castleman’s Disease in A Child – Case Report & Literature Review

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The most prominent sites of Castleman’s disease (CD) are the thorax (mediastinum or lung hilum) and abdomen. It rarely occurs in the cervical area and only one case has been reported in the posterior triangle of the neck. We report a new case of cervical posterior triangle CD in a child. A 9-year-old boy presented with an indolent mass in the posterior triangle (level V) on the left side of his neck for more than six months. Complete excision was undertaken and the histopathological diagnosis was CD of the hyaline-vascular type. At the 3-year follow up, there were no signs of recurrence. The etiologies of persistent cervical lymphadenopathy in children vary. Though it is very rare, we should keep in mind that CD is possible. In addition, CD can be either localized (unicentric) or diffuse (multicentric). The treatment and prognosis of these two are quite different. Therefore, for any case of CD, systemic evaluation is recommended for a precise diagnosis and proper management. (Chang Gung Med J 2011;34:435-9)

Key words: angiofollicular lymph node hyperplasia, castleman’s disease, neck mass

CASE REPORT

A 9-year-old boy presented with a single, painless mass on the left side of his neck for more than 6 months. He denied other associated comorbidity or systemic disease. Under physical examination, a firm, movable neck mass about 2 x 2 cm was noted in the left posterior triangle (level V) of his neck. The skin over the mass was normal, and no compression of the surrounding structures was noted. Routine chemistry and complete blood count data were all within normal limits. Computed tomographic (CT) images of the neck with contrast medium enhancement showed a 1.5 x 2 cm well-defined mass in the left cervical posterior triangle (Fig. 1). Complete excision of the neck mass was performed.

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under general anesthesia. A histopathological diagnosis of CD, HV type (Fig. 2), was made. Since CD can be unicentric or multicentric, a whole body physical examination and chest and abdominal CT were done to exclude possible ignored lesions. However, no further masses or any comorbidity was found. The patient was followed up in our department for 3 years with no recurrence.

**DISCUSSION**

CD is a rare disorder of the lymphoid tissue. Since the first report in 1954, just over 500 cases have been reported, of which 57 were localized to the cervical region. Patient ages ranged from two months to 76 years, with a peak incidence in the second and third decades. No predilection for either gender noted. Cervical CD is very uncommon in children, with only 24 cases reported previously in the literature, comprising only 23% of CD in children. Only one case of cervical posterior triangle CD has been reported previously.

The etiology of CD remains unclear, but there is evidence that certain viral infections or a chronic inflammatory reaction will induce the disease. It has attracted attention because of its association with the human immunodeficiency virus and human herpesvirus 8.

The unicentric HV type is the most common variant encountered clinically. By definition, a single node or chain of lymph nodes is involved. It usually presents asymptptomatically except for associated symptoms induced by the mass effect. Histologically, it is characterized by prominent proliferation of small, hyalinized follicles with marked interfollicular vascular proliferation. The follicles are round and variable in size, surrounded by a cuff of small lymphocytes arranged in concentric “onion skin” layers, with germinal centers frequently demonstrating atrophy with radically penetrating blood vessels.

Under unenhanced CT imaging, the unicentric HV type usually expresses a homogenous mass of soft tissue attenuation, and will express a heterogenous mass under enhancement. Calcification is uncommon, occurring in 5-10% of cases, and is typically coarse and central in the location. Strip enhancement can be found after a bolus injection of contrast material and rim-like enhancement can be found on a 5-minute -delayed scan. In magnetic resonance imaging, T1-weighted images are typically heterogenous and of increased signal intensity compared with skeletal muscle. They become markedly hyperintense on T2-weighted images.

The PC type appears in only 10% of unicentric disease. Unlike the HV type, approximately 50% of patients have systemic findings of anemia, an elevated erythrocyte sedimentation rate, hypergammaglobulinemia and bone marrow plasmacytosis. Histologically, continuous sheets of dense plasma cell infiltration and less vascular interfollicular stroma surrounding the germinal centers are prominent features of the PC type.
Both the HV and PC types of unicentric CD are amenable to complete resection by surgery for a cure. If surgery is not possible, radiotherapy can be considered. The prognosis of unicentric CD is good for both the HV and PC types. The recurrence rate of unicentric CD is very low. According to a recent study, no matter how deep or superficial the location, patients with unicentric CD had no signs of recurrence after a mean follow-up of 56 months after complete resection. Another study showed a 100% 5-year control rate for the HV type of unicentric CD. In addition, one case report showed a good outcome even after a second surgical intervention.

The multicentric type is a systemic disease with disseminated lymphadenopathy, hepatosplenomegaly and constitutional symptoms. Histologically, it is similar to the unicentric PC type, but it occurs in multiple locations with a larger involved range than the unicentric type. On CT images, diffuse lymphadenopathy involving multiple areas can be noted. A homogenous mass presentation can be found on unenhanced CT. Most cases of the PC type will be enhanced by iodinated contrast material but the degree of enhancement may be less and vary more than the HV type.

The most common therapy for multicentric CD is high dose steroids, chemotherapy alone, or combined therapy. Because of the nature of disseminated lymphadenopathy, complete resection is rarely possible. Radiotherapy is another choice but only 23% of patients have favorable responses. Alternative therapies including anti-IL-6 monoclonal antibodies interferon-alpha, antiviral therapy and high-dose melphalan with autologous bone marrow transplantation have been reported.

The prognosis of multicentric disease is poor, with a median survival range from 14 to 30 months. The main causes of mortality are infections which cause sepsis, multi-organ system failure, and malignancies. According to some studies, multicentric disease may have a risk of progressing to lymphoma and Kaposi’s sarcoma.

Conclusion

CD is an uncommon disease of unknown etiology that induces reactive lymph node hyperplasia. Histopathological evaluation is the only way to make a definite diagnosis. We report a new case of unicentric CD in a child whose clinical manifestations were compatible with previous studies except for the unusual location. It is worth mentioning that there are many causes of persistent cervical lymphadenopathy in children. Most are not significant, but some are life-threatening. Although it is very rare, CD should be kept in mind as a possible diagnosis. In addition, we want to emphasize that CD could be either localized (unicentric) or diffuse (multicentric). Multicentric CD is also likely to be misdiagnosed as unicentric initially. Therefore, all patients with proven CD should have a further systemic survey for a precise diagnosis and proper management.

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發生於孩童之頸部後三角卡所門氏病 (Castleman’s disease) —
病例報告及文獻回顧

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卡所門氏病 (Castleman’s disease) 乃一罕見的良性腫瘤疾病，最常出現的部位為縱膈腔及
後腹壁區，出現在頸部區域則較為少見，其中以 level III 為頸部之中最常出現的位置，而於過
去的文獻中，腫瘤出現於頸部後三角區 (level V) 的病例，僅零星個案曾被發表。我們報告一
例九歲男童出現頸部後三角區之腫瘤，經病理切片檢查確診為血管型之卡所門氏病，術前之
理學檢查、血液檢查及術後之胸部電腦斷層均未發現其他病灶，經三年的追蹤並無復發的
情形。兒童頸部腫塊的病因非常多彩，而卡所門氏病則是其中非常罕見的診斷之一，雖然大
多數卡所門氏病皆為侷限型，但約 10% 的病人為廣泛型，且兩者的治療方式及預後有著極大
的差異，因此我們建議對於任何卡所門氏病之患者，均應安排全身性之檢查以得到適當的診
斷及治療。本篇回顧近期文獻，並報告卡所門氏病的類型、臨床症狀、病理特徵、治療方式
及預後不同之處。(長庚醫誌 2011;34:435-9)

關鍵詞：巨大淋巴結增殖，卡所門氏病，頸部腫瘤

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