Castleman Disease in the Pediatric Neck: Case Report and Literature Review

Mindy R. Rabinowitz, MD
Thomas Jefferson University Hospital, Mindy.Rabinowitz@jefferson.edu

Jessica R. Levi, MD
Thomas Jefferson University Hospital

Katrina Conard, MD
Nemours/Alfred I. duPont Hospital for Children, Wilmington, DE

Udayan Shah, MD
Nemours/Alfred I. duPont Hospital for Children, Wilmington, DE

Let us know how access to this document benefits you
Follow this and additional works at: http://jdc.jefferson.edu/otograndrounds

Recommended Citation

This Article is brought to you for free and open access by the Jefferson Digital Commons. The Jefferson Digital Commons is a service of Thomas Jefferson University’s Center for Teaching and Learning (CTL). The Commons is a showcase for Jefferson books and journals, peer-reviewed scholarly publications, unique historical collections from the University archives, and teaching tools. The Jefferson Digital Commons allows researchers and interested readers anywhere in the world to learn about and keep up to date with Jefferson scholarship. This article has been accepted for inclusion in Department of Otolaryngology - Head and Neck Surgery Faculty, Presentations and Grand Rounds by an authorized administrator of the Jefferson Digital Commons. For more information, please contact: JeffersonDigitalCommons@jefferson.edu.
**ABSTRACT**

Castelman disease (CD) was first described by Benjamin Castelman in 1954. Since that time, CD has become better known in literature as a lymphoproliferative disorder of unknown etiology. CD can occur anywhere throughout the lymphatic system. The most common sites include the mediastinum (60%), neck (14%), abdomen (11%), and axilla (4%). While the underlying etiology is unknown, several hypotheses have been suggested. One theory postulates that the disease represents a reaction to a chronic viral antigenic stimulation. Some studies cite a role for interleukin-6 (IL-6). CD is rare in the pediatric population, although exact prevalence rates are not known. In children, CD 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%). In children, the chest (33%) remains the most common site of disease, followed by the abdomen (30%), neck (23%) and axilla (7%).

**INTRODUCTION**

A 13-year-old female presented to our institution with a tender right neck mass that appeared suddenly. It had been present for six weeks, during which time the mass had not changed in size but did cause mild pain with head movement to the right. She had no significant previous medical history. Exam revealed a 4 x 6 cm mass deep to the lower half of the sternocleidomastoid muscle on the right. It was non-tender to palpation with no overlying skin changes. Routine laboratory tests were within normal limits. Titers for toxoplasma, cytomegalovirus (CMV), and Bartonella were negative. Epstein–Barr virus (EBV) IgG titer were elevated. Chest radiograph was within normal limits. Magnetic resonance imaging (MRI) revealed a well-defined, right-sided level III mass measuring 1.8 x 3.0 x 4.2 cm. It was bright on T2- and intermediate to slightly brighter than muscle on T1-weighted imaging (Figure 1). Several small vascular channels were apparent on the lesion by MRI. Several small lymph nodes along the inferior margin of the lesion extending down to the thoracic inlet were also noted. An additional 12 x 6 x 9 mm lesion was noted in the paraspinal musculature. A decision was made not to pursue this lesion given its small size and location, which would be unusual for CD. Fine needle aspiration (FNA) revealed atypical lymphoid proliferation. After discussion with the patient and family regarding the options, she was taken to the operating room, where a complete surgical excision of the mass was performed. Histologically, the nodal mantle zone showed concentric rings with an onion-skin appearance. Puncturing blood vessels were frequently seen in the follicles ("lollipop" feature) (Figure 2). The inner vascular areas showed prominent hyalinized venules. Based on these findings, the postoperative histopathological diagnosis was HV-CD. At three months’ follow-up, she was doing well with no signs of recurrence.

**CASE REPORT**

**MATERIALS AND METHODS**

Online medical journal databases were used for data collection. “Castleman’s disease” in combination with “neck”, “cervical”, and “pediatric” were keywords used for searching the PubMed database. Only patients aged 18 and younger were included for analysis. After excluding reports on CD in other locations (ie, non-neck sites), 18 published papers were found, comprising 29 total reported cases of pediatric cervical CD (Table 1). The earliest case report published was in 1991 and the latest in 2012. In addition, one patient was diagnosed and treated at our institution. This patient was also included and brought our final patient count to 30 cases. This study was IRB exempt. All diagnoses of CD were based on histopathology.

**DISCUSSION**

Cervical pediatric CD is rare. It most commonly presents as an asymptomatic or slowly enlarging left mass. Imaging characteristics are often non-specific and do not aid in the diagnosis. Imaging is important in excluding other diagnoses and to allow for presurgical planning. The specific lab abnormality in cervical pediatric patients, nor is FNA diagnostic. Excision is ultimately diagnostic and therapeutic, and when presenting in the pediatric neck, the diagnosis is likely HV-CD, which holds a favorable prognosis.

**REFERENCES**