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
Part of the Otolaryngology Commons

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.
Castelman Disease in the Pediatric Neck: Case Report and Literature Review

Mindy R. Rabinowitz, MD; Jessica R. Levi, MD; Katrina Conard, MD; Udayan Shah, MD

1 Department of Otolaryngology-Head & Neck Surgery and 2 Department of Pediatrics, Thomas Jefferson University Hospital, Philadelphia, PA; 3 Department of Pathology and 4 Division of Otolaryngology, Nemours/Alfred I. duPont Hospital for Children, Wilmington, DE. Work performed at Nemours/Alfred I. duPont Hospital for Children.

ABSTRACT

In adults, the most common locations for CD include the chest (60%), neck (14%), and abdomen (11%). This report describes a unique presentation of pediatric CD involving the neck. A 13-year-old female presented to our institution with a tender right neck mass. MRI revealed a large mass in the prevertebral space with invasion of the sternocleidomastoid muscle. The mass was biopsied and histopathology confirmed Castleman disease. This is the first report of Castleman disease involving the neck in a paediatric population.

INTRODUCTION

Castleman disease (CD) is a rare disorder that may be either solitary (unicentric) or multicentric. Solitary CD is more common in adults, whereas multicentric CD is more common in children. While Castleman disease is rare in the pediatric population, the exact prevalence rates are unknown. The underlying etiology is unknown, and several hypotheses have been suggested, including infectious and autoimmune mechanisms.

CASE REPORT

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. Tilters 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 immediately to slightly brighter than muscle on T1 weighted imaging (Figure 1). Several small vascular channels were apparent on the lesion by MRI. Several 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 unsuitable for CD. Fine needle aspiration (FNA) revealed atypical lymphoid proliferation. After discussion with the patient and family, they elected to undergo complete excision. 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.

DISCUSSION

In adults, the most common locations for CD include the chest (60%), neck (14%), and abdomen (11%). In children, the chest (33%) remains the most common site of disease, followed by the abdomen (30%), neck (23%) and axilla (7%).4,5 Data are currently inconclusive as to the most common neck level for pediatric CD. In our literature analysis, level V was the most common location, representing 25% of lesions.

All of the evaluated children had unicentric masses. While no multicentric disease was found, it is important to note that only 17% (5 of 30) of children in this literature review received full body work-up to rule out this possibility. Therefore, it is difficult to make this conclusion definitively.

Radiographic imaging is non-specific.6 CT was the most common modality used for neck mass work-up in this analysis (47%); however, the results argue that CT is no more specific for diagnosing CD than any other modality. This is supported by other studies that have found that CT is no more specific for diagnosing CD than any other modality. CT is primarily used to rule out other diagnoses and to aid in surgical planning.

Online medical journal databases were searched 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. After discussing 29 total reported cases of pediatric cervical CD (Table 1). The earliest case report was published in 1991 and the latest in 2012.5,9 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.

MATERIALS AND METHODS

Online medical journal databases were searched 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. After discussing 29 total reported cases of pediatric cervical CD (Table 1). The earliest case report was published in 1991 and the latest in 2012.5,9 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.

CONCLUSIONS

Cervical pediatric CD is rare. It most commonly presents as an asymptomatic or slowly enlarging V mass. Imaging characteristics are often non-specific and do not aid in the diagnosis. Diagnosis is important in excluding other diagnoses and for a timely treatment plan. In children, the neck is a particularly relevant location to evaluate. Future studies should focus on the clinical characteristics of pediatric Castleman disease.

REFERENCES