Unicentric Castleman Disease in the Temporal Region of a Pediatric Patient

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Introduction

Castleman disease (CD) encompasses a group of rare lymphoproliferative disorders which share common histopathologic features. The most common subtype is unicentric CD (UCD) which has unclear etiology and involves a singular region of enlarged lymph nodes. UCD is generally asymptomatic and tends to occur in adults. CD can occur in any lymph node; however, it has a predilection for the mediastinum, and rarely presents on the face.[1] Here we report a case of symptomatic UCD presenting as a superficial mass in the temporal region of a pediatric patient.

Case Presentation

An 11-year-old female presented to her primary care provider with an enlarging soft lump on the right temple. Associated symptoms included periodic tenderness and fever for two weeks prior to presentation. An MRI revealed a well-circumscribed 3.4 cm x 1.4 cm x 3 cm extracranial soft tissue lesion lateral to the right temporalis muscle with vascular flow voids indicative of increased vascularity. She was referred to our dermatological surgery center for excisional biopsy. A crescentic excision was performed, oriented along the hairline at the border of the lateral forehead cosmetic subunit, and the resultant defect was closed in a layered fashion following the rule of halves (Figure 1 & 2).

Histological analysis of the excised specimen revealed lymphoid tissue with atretic and hyalinized germinal centers, concentric laminations resulting in an “onion-skin appearance,” and penetrating hyalinized arterioles (Figure 3 & 4).

Immunohistochemistry demonstrated CD20 and PAX-5 positive B cells within the germinal centers, as well as CD21 positive follicular dendritic cells forming a laminated meshwork and occasionally merging with neighboring germinal centers. These findings, along with the absence of prominent plasmacytosis, atypical lymphocytes, and large or transformed lymphoid populations are consistent with the histologic features of UCD, and lead to the patient’s ultimate diagnosis.

Figure 1. Surgical defect following excisional biopsy

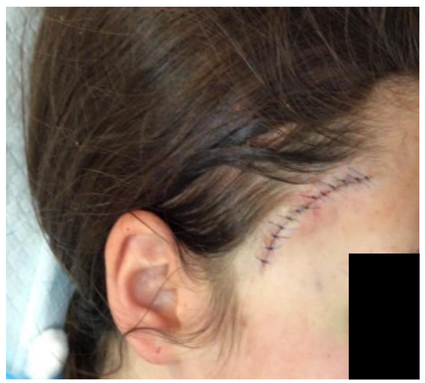

Figure 2. Crescentic closure along the lateral forehead cosmetic subunit

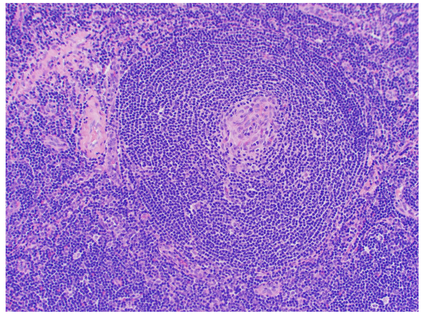

Figure 3. Lymphoid tissue with atretic and hyalinized germinal centers and concentric lamellations resulting in an “onion-skin” appearance

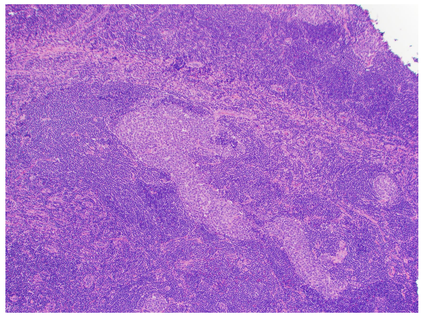

Figure 4. Lymphoid tissue and penetrating hyalinized arterioles

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Discussion

Castleman disease, also called angiofollicular lymph node hyperplasia, is a group of rare lymphoproliferative disorders with common histopathologic features. CD can be categorized into two groups: unicentric and multicentric. UCD is localized, involves a single region of enlarged lymph nodes, and is more common than multicentric CD (MCD).[2] MCD affects multiple lymph node regions and usually presents with systemic symptoms, such as fever and fatigue. MCD can further be classified as either human herpesvirus 8 (HHV-8) positive or HHV8-negative.[2] There are three major histological subtypes: hyaline-vascular CD (HV-CD), plasma cell CD (PC-CD) and a mixed histopathologic subtype. Over half of UCD cases demonstrate the hyaline-vascular CD morphology, whereas MCD is associated with the plasma cell variant.[3]

Approximately 6,600-7,7000 new cases of Castleman Disease are diagnosed each year in the United States, however, only a small fraction of these cases occur in the pediatric population.[4] Since 2011, there have only been 100 published cases in pediatric patients, of which UCD is the most common. The involved lymph node is usually large with a median diameter of 6-7 cm and identified as a singular mediastinal mass.[5] It is crucial to accurately diagnose the subtype of CD since each group requires different treatment strategies.

A complete resection of the lymphoid tissue is almost always curative for UCD, and surgical excision is considered the gold standard for treatment.[6] There have been two other case reports of CD presenting in the temporal region. This case is unique given the presentation of symptomatic temporal UCD in a pediatric patient.[1,3] It is our hope that by sharing this case, dermatologists will continue to be aware that although uncommon, the development of CD is possible in the temporal region. Although the exact etiology of CD remains to be determined, this case serves as a reminder that when confronted with a solitary mass of the temporal region, further workup is required, especially when systemic symptoms are present.