Dermatology Practical & Conceptual

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 with common histopathologic features. The most common subtype is unicentric CD (UCD) which involves a single region of enlarged lymph nodes. CD can occur in any lymph node; however, CD has a predilection for the mediastinum, and rarely presents on the face [1]. Here we report a case of UCD presenting in the temporal region of a pediatric patient.

Case Presentation

An 11-year-old female presented for excisional biopsy of an enlarging mass on the right temple which had been present for 17 months and was occasionally tender. An MRI revealed a well-circumscribed 3.4 cm x 1.4 cm x 3 cm soft tissue mass lateral to the temporalis muscle.

Histological analysis of the specimen revealed lymphoid tissue with atretic and hyalinized germinal centers, concentric laminations resulting in an "onion-skin appearance," and penetrating hyalinized arterioles (Figure 1). Immunohistochemistry demonstrated CD20 and PAX-5 positive B cells

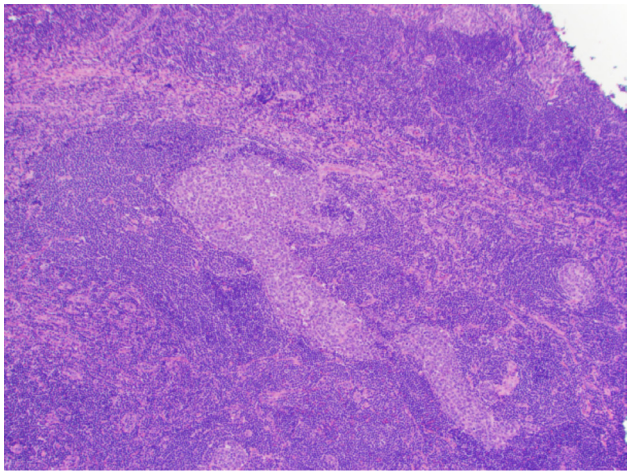

Figure 1. Lymphoid tissue with atretic and hyalinized germinal centers, concentric laminations resulting in an "onion-skin appearance," and penetrating hyalinized arterioles (H&E, 40x).

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 in a single region, along with the absence of prominent plasmacytosis, atypical lymphocytes, and large or transformed lymphoid populations are consistent with the features of UCD.

Conclusions

CD is categorized into two groups: unicentric and multicentric. UCD involves a single region of enlarged lymph nodes and is more common than multicentric CD (MCD). MCD affects multiple lymph node regions and usually presents with systemic symptoms or laboratory abnormalities demonstrating chronic inflammation such as elevated C-reactive protein [2]. There are three major histological subtypes of CD: 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 more often demonstrates the plasma cell variant [3].

UCD is typically asymptomatic and often discovered as an incidental finding on imaging or physical examination; however, it can also present with mass effect on neighboring structures [3,4]. UCD in the pediatric population is most commonly found within the mediastinum [4].

A complete resection of the lymphoid tissue is almost always curative for UCD. It is unknown if UCD in the temporal region carries a distinct prognosis from other locations, however, the prognosis following complete resection of UCD affecting a peripheral lymph node is significantly better than UCD in the chest or abdominal cavities. In a systematic review of 278 UCD cases, 249 had surgical resection alone, and 13 had combination surgical and immunosuppressive treatments with a 5-year disease free recurrence rate of approximately 81% and a 10-year survival rate of approximately 95%. Continued follow-up is essential to monitor for recurrences. Treatment of MCD is more complex and the prognosis is generally worse with a 10-year survival rate of approximately 67% [5].

There have been two other case reports of CD presenting in the temporal region [1,3]. This case is unique given the presentation of temporal UCD in a pediatric patient. Dermatologists should be aware that CD may develop in this region in both children and adults.

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