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654.MGUS, AMYLOIDOSIS AND OTHER NON-MYELOMA PLASMA CELL DYSCRASIAS: CLINICAL AND EPIDEMIOLOGICAL

A Rare Case of Unilateral Lymphadenopathy: Unicentric Castleman Disease

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Background

Castleman Disease (CD) involves a group of disorders characterized by an enlargement of lymphoid follicles with capillary proliferation. The disorders are largely separated into unicentric (UCD) or multicentric (MCD) depending on the number of involved lymph nodes. MCD has been linked to HHV8 or immunosuppression secondary to HIV, while UCD has been thought to be driven by a clonal neoplastic process, likely from the follicular dendritic cell.

Case Presentation

A 14-year-old female with ADHD, OCD, and autism spectrum disorder (ASD) presents to clinic with a 2-year history of a right-sided neck mass. There was no history of trauma or illness around the time of the mass being noted and the mass has not changed significantly in size for the past two years. An excisional biopsy was attempted but, due to excessive vascularity around the mass, was ultimately deferred. Following subsequent MRI and CT scans, a core needle biopsy was performed, revealing lymphoid tissue with regressed follicles and focal hyalinization without signs of malignancy. PET/CT scans at this time showed increased metabolic activity confined to the right-sided neck mass without any increased metabolic activity in the other lymph nodes of the head and neck. Eventually, the 7 cm diameter mass was excised; pathology studies revealed distorted architecture with hyaline deposits in the germinal centers and sclerotic blood vessels penetrating the germinal centers consistent with the hypervascular variant of Castleman Disease. Pre- and post-operative labs such as the C-reactive protein, immunoglobulin counts, and protein electrophoresis panels were all within normal limits. Furthermore, post-operative PET/CT scans showed no metabolically active foci indicating that the lesion was indeed unicentric Castleman Disease. After the excision of the mass, the patient continues to do well without any further surgeries or complications.

Discussion

Differentiation between UCD and MCD requires imaging, often with a CT scan, to establish the number and locality of the affected lymph nodes. UCD often presents with one bulky lymph node or multiple enlarged lymph nodes within the same region, whereas MCD tends to be smaller lymph nodes spread over multiple regions. MCD is also more often associated with B symptoms and abnormal lab values such as elevated CRP or ESR along with abnormal protein levels. Other subtypes exist, thought to be driven primarily by cytokines like VEGF and IL-12 similar to other neoplasms. The hypervascular type, more commonly associated with UCD, consists of fibrous bands running through lymphoid follicles with regressed germinal centers while the plasmacytic type, found more often in MCD, consists of sheets of plasma cells and hyperplastic germinal centers. Lastly, mixed type has features of both the main types. Preferred treatment for resectable UCD is surgery with treatments for unresectable UCD involving observation (if asymptomatic) or a combination of rituximab and steroids with anti-IL6 monoclonal antibody treatment (such as tocilizumab) followed by surgery or radiation therapy. Similarly, MCD is often treated with rituximab, steroids, and tocilizumab.

Conclusion

Due to the presence or absence of other signs and symptoms, Castleman Disease is often difficult to diagnose without a biopsy. Further studies into the etiologies of CD, including possible associations with other neoplastic or immunosuppressed states, are needed to develop better criteria for diagnosing the disease. Developing standardized patient registries with information on disease progression as well as proper follow-up for patients after treatment are vital to diagnosing, treating, and providing surveillance for patients with Castleman Disease.

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