Castleman Disease of Adrenal Masquerades as Adrenal Carcinoma; Review Article on Current Treatment Modalities

Adrenal Karsinom Olarak Adrenal Maskeli Castleman Hastalığı; Mevcut Tedavi Yöntemleri Üzerine Makale Derlemesi

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ABSTRACT

Castleman disease is a rare lymphoproliferative disorder. Only few cases reported worldwide since first describe in 1956. Only 2 percent of CD involve pararenal location. We report on a 16 years old lady with solitary Castleman disease of hyaline vascular type, involving right adrenal. Patient underwent right open adrenalectomy and no evidence of disease recurrence in 1 year follow up after surgery. In addition, we also discuss on the pathogenesis of the disease, clinical features and option of treatment.

Key Words: Castleman disease, adrenal, unicentric

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ÖZET

Castleman hastalığı nadir görülen bir lenfoproliferatif hastalıktır. İlk kez 1956'da tanımlandığından bu yana dünya çapında rapor edilen çok az vaka vardır. Bu yazıda, hyalin vasküler tipte soliter Castleman hastalığı olan ve sağ adrenal bulunan 16 yaşında bir kadın hasta sunuldu. Hastaya sağ açık adrenalektomi uygulandı ve cerrahi sonrası 1 yıllık takipte hastalık nüksü izlenmedi. Ek olarak, hastalığın patogenezi, klinik özellikleri ve tedavi seçeneği hakkında da tartışıyoruz.

Anahtar Sözcükler: Castleman hastalığı, adrenal, tek merkezli

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INTRODUCTION

Castleman disease or called as atypical lympho-proliferative disease or proliferative disease with uncertain malignant potential was first described by Castleman, Iverson and Menendez in 1956 (1) is a rare and heterogeneous disorder with few case series study have found a new knowledge of its disease natural history, etiology, outcome as well as the treatment option.

It has been suggested that anomalous proliferation of B lymphocytes in response to infectious or chronic stimuli in individuals with a genetic predisposition, autoimmunity or immunodeficiency disorder. We report on our first experience in surgical management of this uncommon condition and literature review on current treatment modalities on this disease.

CASE REPORT

A 16 year-old Javanese had suffered from worsening recurrent right hypochondric pain for 1 year duration, stabbing in nature, not associated with meal and usually it resolved spontaneously. Clinically she is morbidly obese with BMI of 40.2 with presence of striae on the abdomen and fullness of bilateral supraclavicular fat pad. Otherwise no other systemic symptoms and physical examination was unremarkable. Laboratory findings for renal, liver function and full blood count were within normal range. Sonography of abdomen shows no evidence of cholelithiasis or billiary obstruction. Noted right suprarenal mass with small right ovarian cyst, thus proceeded with CT abdomen and pelvis which reveal homogenous enhancing mass measuring 5.8 x 8.0 x 6.0 cm on right suprarenal adrenal mass, given the impression of likely an adenoma with mild central hypodence area to suggest necrosis. Mass was encapsulated, receiving blood supply direct from the abdominal aorta. . The left ovarian dermoid cyst measured 4.8 x 4.5 cm. CT adrenal with adrenal protocol shows relative percentage of washout of less than 40 percent, suggestive of malignant adrenal tumor. Further work out from endocrine team suggested that it was a non-functional tumor. Patient was then underwent right adrenalectomy and recovered well post-operative. Histopathology examination of the tumor shows a lymphoid tissue composed of scattered lymphoid follicles with surrounding mature interfollicular lymphocytes with prominent vascular proliferation (Figure 1). There were marked penetrating vessels and hyalinization seen traversed within the atretic germinal centers (Figure 2). Immunohistochemical study showed maintenance of non-neoplastic architecture of lymphoid follicles evidence by the presence of a network of CD21 positive dendritic follicular cells (figure 3). The mantle zones are thickened with multiple concentric layers of lymphocytes giving an onion skin appearance (figure 4). No mitosis or nuclear atypia noted giving a diagnosis of Castleman Disease (Giant lymph node hyperplasia), hyaline-vascular-type. Patient was followed up under surgery clinic for almost one year post right adrenalectomy and does not has any evidence of recurrence.

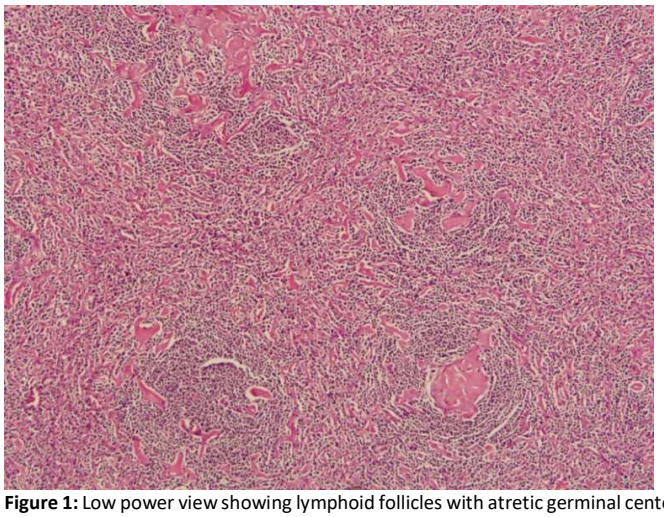

Figure 1: Low power view showing lymphoid follicles with atretic germinal center and prominent vascular proliferation. (H&E, original magnification x10)

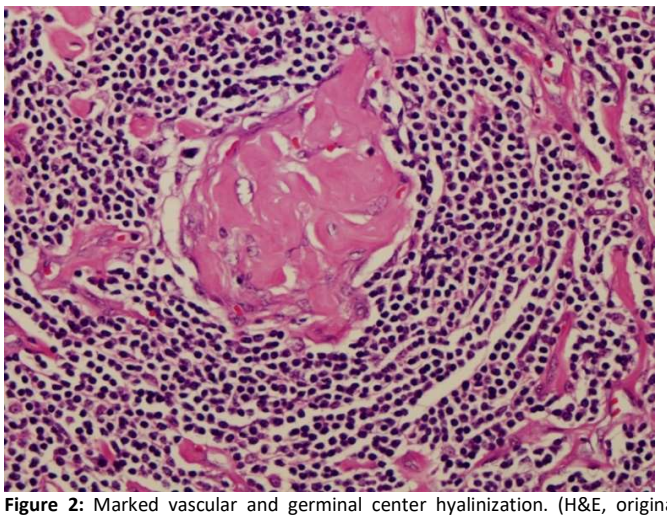

Figure 2: Marked vascular and germinal center hyalinization. (H&E, original magnification x20)

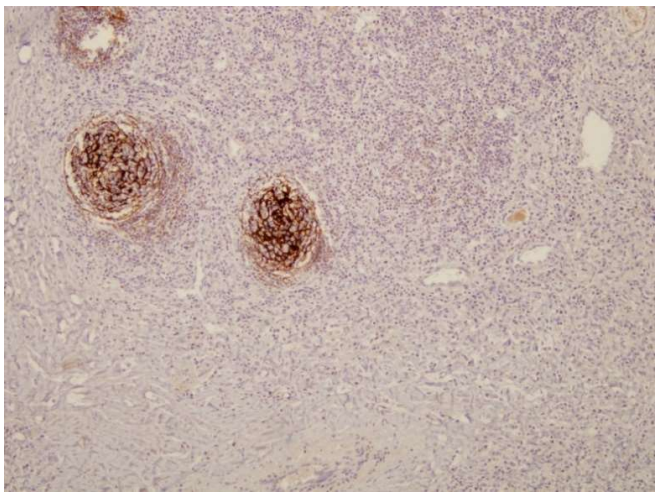

Figure 3: Presence of a network of CD21 positive dendritic follicular cells. (Immunohistochemical stain, original magnification x4)

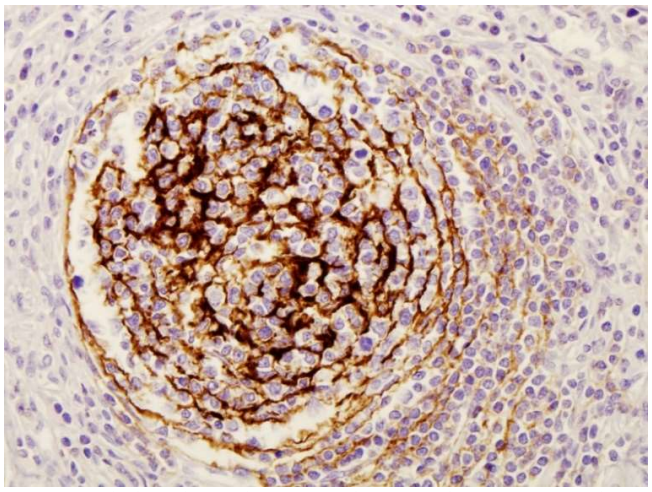

Figure 4: Thickened mantle zone with onion skin appearance layers of lymphocytes. (Immunohistochemical stain, original magnification x40)

DISCUSSION

CD is a poorly understood disease that creates both diagnostic and therapeutic dilemmas for the treating physician . CD especially MCD can be a great mimicker of many other benign or malignant neoplasm. Due to presence of masses and lymphadenopathy, this will raise a suspicion of malignant tumor of lymphoma or metastatic tumor or even a benign condition of infectious and inflammatory disease (2). It is difficult to distinguished CD from other tumors if based on their clinical manifestations or radiological findings (3). Some reactive cause of lymphadenopathy may be indistinguishable from CD. A biopsy is required in particular to differentiate between lymphoma and disseminated CD. Proper histological evaluation is needed in order not to misdiagnose such cases.

Generally it can be divided into two type, Unicentric Castleman disease (UCD) is localized disease and carries an excellent prognosis, whereas multicentric Castleman disease (MCD) is a systemic disease with poorer prognosis. It occur most commonly in the setting of HIV infection and is associated with human herpesvirus 8.(4). The therapeutic landscape for its management continues to evolve.

Surgical resection is recommended for patients with the unicentric variant of CD and it is a curative measure for hyaline-vascular or hyaline-vascular/plasma cell type of UCD. Partial resection, radiotherapy, or observation alone may avoid the need for excessively aggressive therapy. Patients with multicentric disease usually do not benefit from surgical management and should be candidates for multimodality therapy, the nature of which has yet to be defined (5). Radiation therapy has been used with varied success in patients who are poor surgical candidates or in those with unresectable lesions. Long term follow-up is necessary with regard to malignant sequelae. The role of surgery in multicentric disease is limited and should not be considered a realistic treatment option. Systemic therapy in the form of steroids or single- or multiple-drug chemotherapies have all been used with varied success. However, there is no evidence for one approach being more consistently effective. This study also highlights a group of patients with multicentric disease and neuropathy for whom prognosis is poor and investigational treatment is justified.

A better understanding of both the pathogenesis and the natural history of this disorder will lead to improvement over the current modalities available for treatment (5).

CONCLUSION

Castleman's disease should be considered as a differential diagnosis in case well-defined non-functional tumor in a patient without risk factors for malignancy or infection. Surgical excision is required for the diagnosis. Unicentric disease of hyaline vascular variant is associated with complete recovery after surgical resection.

Conflict of interest

No conflict of interest was declared by the authors.

REFERENCES

1. Costleman B, Towne V. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises: case 40011. N Engl J Med. 1954;250:26-30.
2. Luisa R, Fabiana F, Antonio I, Antonio M. Castleman Disease in a Patient with Common Variable Immunodeficiency. Hindawi Case Reports in Immunology, vol. 2019, Article ID 5476383, 5 pages, 2019.
3. Xie Y, Zhao Y, Ji ZG, Li HZ, Liu GH, Mao QZ. Castleman's Disease: A Rare Mass in the Pararenal Retroperitoneum that Mimics Other Tumors. Chinese Medical Journal (Engl). 2017;130:2126–7.
4. Soumerai JD, Sohani AR, Abramson JS. Diagnosis and management of Castleman disease. Cancer control. 2014;21:266-78.
5. Bowne WB, Lewis JJ, Filippa DA, Niesvizky R, Brooks AD, Burt ME, et al. The management of unicentric and multicentric Castleman's disease: a report of 16 cases and a review of the literature. Cancer: Interdisciplinary International Journal of the American Cancer Society. 1999;85:706-17.