

Castleman's Disease Clinical Case and Literature Review

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Abstract

Case Report

Castleman's disease or angiofollicular lymphoid hyperplasia is a rare condition of uncertain cause. It poses a diagnostic and therapeutic problem. We report the clinical case of a 58-year-old patient who presents LUTS associated with microscopic hematuria with an abdominopelvic CT scan in favor of a 20mm retroperitoneal tissue lesion at the lower pole of the right kidney which is enhanced by injection of PC posing a differential diagnosis problem with the other retroperitoneal masses. The biopsy under scanner was not performed given the difficult localization of the lesion, it was finally lumpectomy with pathological study that made it possible to make the diagnosis. The immediate and remote post-operative consequences were favorable. Through this clinical case and through a literature review, we analyze the epidemiological, diagnostic and therapeutic aspects of this rare pathology.

Keywords: angiofollicular lymphoid hyperplasia, CT scan, retroperitoneal tissue, scanner.

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INTRODUCTION

Castleman's disease or angiofollicular lymphoid hyperplasia is a rare condition of uncertain cause. It poses a diagnostic and therapeutic problem. In fact, there are two clinical forms: localized unicentric form, pseudotumoral with a good prognosis; and multicentric form associated with more aggressive dysimmunity manifestations [1, 2]. Three histological types have been identified: the type with hyalinized vascularization, the plasma cell type and an intermediate mixed type.

In this article, we report on Castleman's disease localized in a 58-year-old patient and analyze, through a review of the literature, the epidemiological, diagnostic and therapeutic aspects of this rare pathology.

PATIENT AND OBSERVATION

Male DO, 58 years old, history of intestinal obstruction at 14 years, smoking weaned 10 years ago, had consulted for LUTS not improved by alpha-blocker treatment associated with microscopic hematuria on several ECBUs without urinary tract infection, without any associated sign, the whole evolving in a context of apyrexia and conservation of the general state.

The clinical examination is unremarkable. The rectal examination shows a flexible 60g prostate without a nodule. Examination of EMBs and lymph node areas is normal.

Ultrasound shows a 75g prostate with 70ml RPM, with normal sized kidneys without abnormalities.

Abdomino-pelvic computed tomography (01/24/2018) shows 2 normal kidneys with the presence of a 20mm retroperitoneal tissue lesion at the lower pole of the right kidney which is enhanced with PC injection.

The PET scan (02/06/2018) shows a hypermetabolic lesion, with an SUV MAX of 4, as well as a thyroid goiter at the expense of the left lobe (Figure 1 & 2).

A report was carried out (05/02/2018): ACE: 2 µg / l; NSE 15 ng / ml; AFP 1.2 ng / ml; HCG <1; Chromogranin A: 123 ng / ml (Chromogranin A down on 03/08/2018 after stopping PPI (109ng / ml)).

Faced with the possibilities of a paraganglioma or a tumor that may be an ectopic pheochromocytoma, urinary metanephrines are negative (02/09/2018).

The biopsy seems difficult given the position of this lesion between the inferior pole of the kidney and the IVC.

The patient underwent a right lumpectomy by lobotomy (03/20/2018). Histological examination shows localized Castelman Benigne GG disease (Figure 3, 4 & 5).

The evolution turned out to be favorable, with no locoregional recurrence or distant recurrence. The follow-up is 1 year.

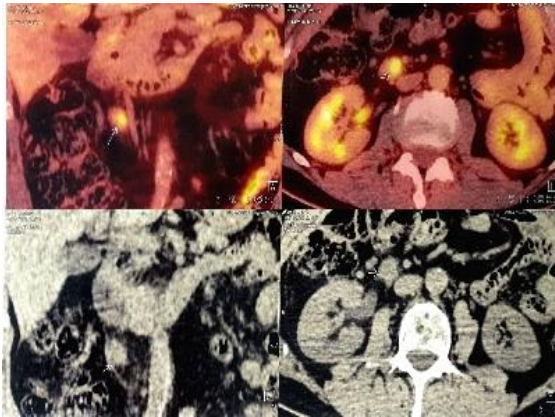


Fig-1

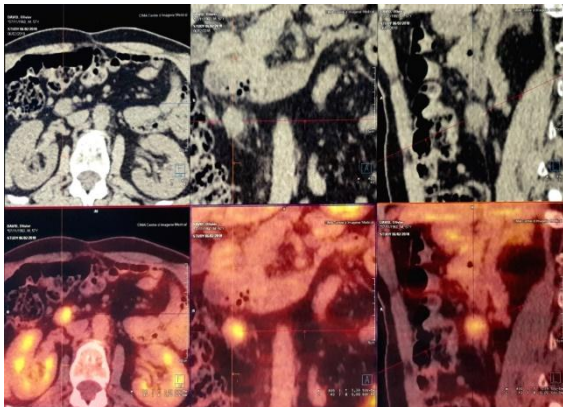


Fig-2

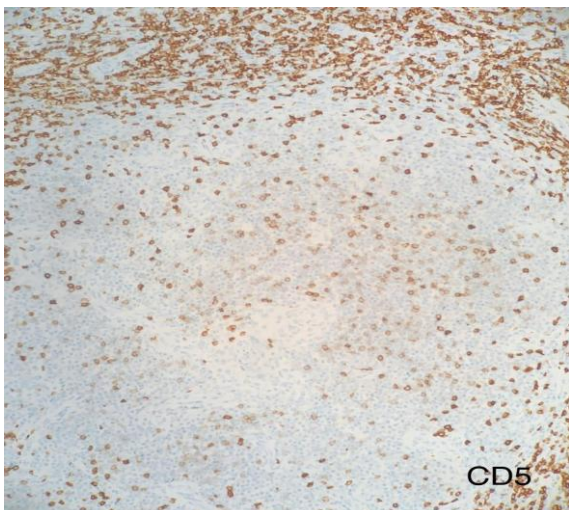


Fig-3

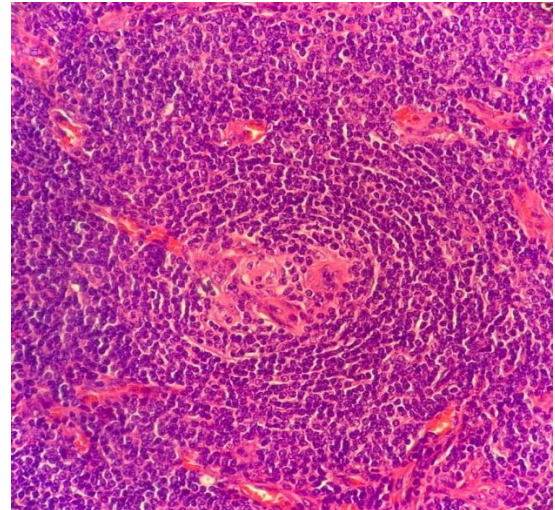


Fig-4

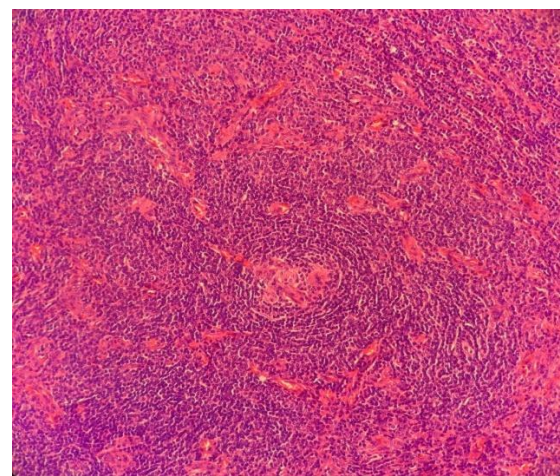


Fig-5

DISCUSSION

Castleman's disease or angiofollicular lymphoid hyperplasia is a rare condition that can be seen at any age. It affects men and women without gender predominance and was first described in 1954 [1].

The etiology of this pathology remains uncertain. However, possible causes include chronic inflammation, immunodeficiency, autoimmunity, tuberculosis, toxoplasmosis, Epstein Barr virus, infection with human herpes virus 8 (HHV8) as well as increased serum levels of interleukin 6 (IL6) [2].

Indeed, there are two clinical forms: localized unicentric form, pseudotumoral with a good prognosis; and multicentric form associated with more aggressive dysimmunity manifestations.

It is symptomatic in less than 10% of cases; revealing either by a tumor syndrome with compression of neighboring organs, or by a systemic inflammatory syndrome with hyperthermia, night sweats and deterioration of the general condition.

Castleman's disease is easily confused with lymphoma or other solid tumors on X-ray examination. The radiological characteristics are not very specific; his diagnosis remains pathological [3]:

Three histological types have been identified in the forms not associated with the HHV-8 virus:

1. The type with hyalinized or Hyper-Vascular (HV) vascularization marked by the importance of the regression of germinal centers, vascular anomalies and anomalies of the network of follicular dendritic cells. It is the most common form of localized CD.
2. The plasma cell type marked by significant interfollicular plasmacytosis, whereas the vascular and follicular dendritic cell abnormalities are modest. It is the most common form of idiopathic multicentric CD.
3. We describe mixed or intermediate forms between HV and PC and in particular a hypervascular form with fibrosis observed rather in Asian patients (TAFRO syndrome).
4. The form associated with the HHV-8 virus is superimposable on the plasma cell form but is distinguished by the presence of "plasma" cells infected with HHV-8 and identifiable by an immunohistochemistry (LANA) technique.

Our observation responds to the unicentric form which is seen in 80 to 90% of cases. The multicentric form, rarer, is seen in 10 to 20% of cases corresponds to the diffusion of lymphatic involvement to multiple lymph node areas, with possible visceral extension. In this form, the clinical and biological inflammatory signs are constant [4, 5].

The first-line treatment for unicentric Castleman's disease is surgical excision; it generally ensures healing when it is complete. Tumor volume reduction can be done for lesions that are difficult to access, followed by monitoring or focal radiotherapy [6-9].

Treatment of multicentric Castleman's disease aims to reduce lymph node mass, treat symptoms, and reverse the inflammatory syndrome and its consequences. Different therapeutic approaches are possible and use corticosteroids, monoclonal antibodies targeting IL-6 or its receptor or even immunosuppressants [10].

For Castleman's disease associated with HHV-8, the current standard of treatment is the use of an anti-B monoclonal antibody, rituximab ®. It is often combined with etoposide which results in rapid improvement of symptoms [10].

Treatment of the associated immune deficiency is fundamental (anti-retroviral treatment for HIV, reduction in immunosuppression during transplants).

Relapses are possible but may again be susceptible to the same treatment [10].

For our patient, the evolution turned out to be favorable, without locoregional recurrence or distant recurrence with a follow-up of 1 year.

CONCLUSION

Castleman's disease is a rare condition of uncertain cause. It poses a diagnostic and therapeutic problem with retroperitoneal solid cancers, hence the need to perform a surgical excision of the lesion with anatomopathological study thus constituting a treatment in the unicentric forms.

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