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Castelman's Disease with Mesenteric Localization

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Abstract **Case Report**

Castelman's disease is a rare affection, which affects the abdomen in 21% of cases. We report the case of a 62-year-old woman, in whom a midline laparotomy allowed excision of a mesenteric mass at the expense of the first jejunal loop, and whose histology is in favor of Castleman's disease.

Keywords: Castelman's disease, Mesentery, surgery.

INTRODUCTION

Castelman's disease was first described by Benjamin Castelman in 1956 [1]. It is an isolated benign or multicentric lymph node involvement with a reserved prognosis, especially in the event of association with other tumoral, inflammatory or infectious pathologies. This atypical lymphoid proliferation presents a clinical and biological diversity requiring a well-established diagnostic approach. Diagnosis is based on anatomopathological examination of the excised specimen or during a lymph node biopsy. Excision surgery remains the best therapeutic means for localized forms and which can be associated with other therapies in multicentric forms, thus allowing healing and avoiding recurrence.

OBSERVATION

A 62-year-old patient was admitted to our surgery department for a mass in the left iliac fossa. Having a history of laparoscopic cholecystectomy and treated lymph node tuberculosis, cured 20 years ago. The history of the disease went back to 2 months of his admission by the appearance of a mass at the level of the left iliac fossa gradually increasing in volume, painless, associated with constipation in a context of conservation of the general state and of apyrexia. The somatic examination found an apyretic patient, hemodynamically and respiratory stable, the abdominal examination effectively objectified a mass of the left iliac fossa, hard, painless, of regular contours measuring approximately 10 cm. There was no clinically palpable adenopathy. An abdominal ultrasound was requested, showing a large heterogeneous tissue mass measuring 100*120*100 mm without specifying its nature. Abdomino-pelvic magnetic resonance imaging was performed showing regular circumferential thickening of the digestive wall, producing the target, oval, well-defined, irregular with clear contours, in T1 and T2 isosignal, restrictive in DW1, homogeneously enhanced after injection of gadolinium measuring 106*145*135 (AP*T*H). Topographically, it encompasses digestive structures without signs of stenosis. Posteriorly, it comes into contact with the external iliac arteries which remain permeable, comes into contact with the vertebral body of L5 without signs of invasion, remains at a distance from the psoas muscles. Anteriorly, comes into contact with the anterior abdominal wall without signs of invasion. At the top, arrives at the height of L4 and includes the mesenteric vessels which remain permeable. Below, comes into contact with the bladder and the sigmoid colon with the fatty border of separation. Laterally it seems to continue with some digestive structures. This appearance was suggestive of lymphoma.

A midline laparotomy was performed, abdominal exploration objectified a solid mass about 15 cm long axis of the first jejunal loop, very adherent to the superior mesenteric vessels with infiltration of the nearby peritoneum.

The release of the superior mesenteric vessels and the detachment of the angle of Treitz allowed the tumor resection taking away the first loops over a length of 40 cm with the making of an end-to-end duodeno-jejunal anastomosis. The post-operative follow-up was simple.

The macroscopic study objectified a mass measuring 15*12*9 cm with a whitish, homogeneous, thin appearance when cut. The histological study showed a lymph node parenchyma with a grossly nodular architecture. The nodules are made up of lymphoid follicles often having onion bulb germinal centers with numerous follicular dendritic cells packed close together and surrounded by a zone of hyperplastic mantle where the cells arrange themselves in single file. The deep cortex is hyperplastic. This description being compatible with Castelman's disease.

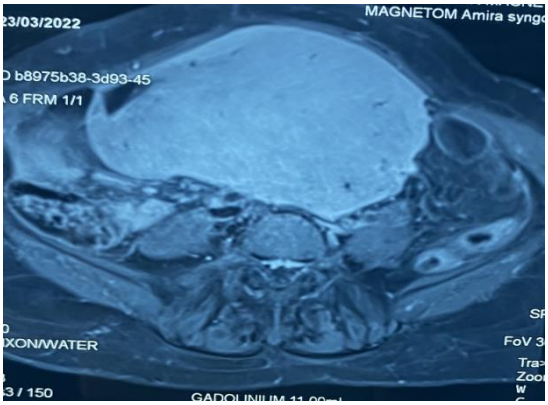

Figure 1: MRI showing a large digestive mass exerting a mass effect on neighboring structures

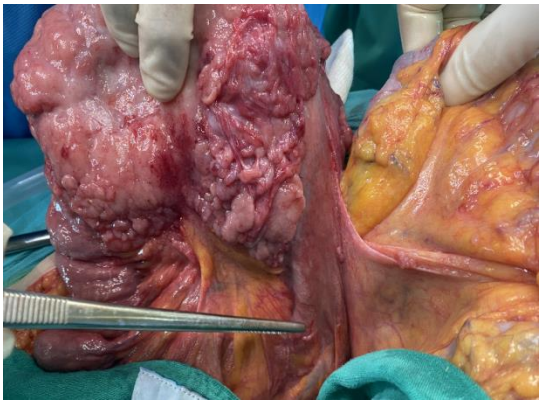

Figure 2: Image showing the mass at the expense of the first jejunal loop

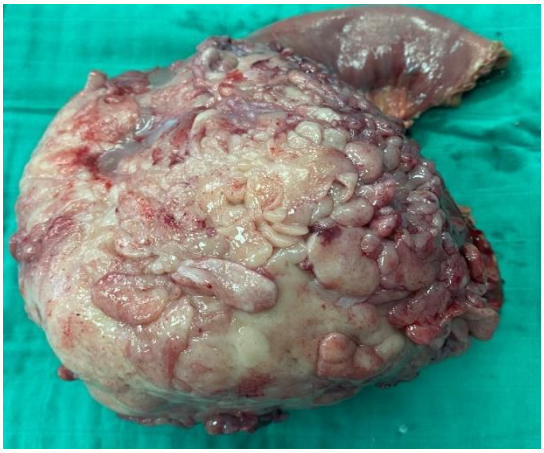

Figure 3: Image showing the mass after resection

DISCUSSION

Castelman's disease is a rare lymphoid lymphoid hyperplasia, which originates in the lymph node chains, especially at the expense of the lymph nodes of the mediastinum with a predominance in the tracheobronchial tree and the pulmonary hiles [1]. Others Localizations of Castelman's disease have been described in the literature, in particular at the abdominal and pelvic level: intraperitoneal in 5.7% of cases (including 3.5% in the mesentery), and retroperitoneal in 6.6% of cases [2]. It can also occur in the axillary and inguinal regions as well as in the orbits, nasopharynx and small intestine [3, 4].

The study of the pathophysiology of this disease has highlighted the key role of interleukin 6, which is produced in large quantities by the lymph nodes, thus triggering a massive inflammatory reaction either locally or systemically if several lymph node sites are affected [5, 6]. It has also been suggested that the herpes virus type 8 (HHV-8) is involved in the genesis of Castelman's disease [7, 8].

Castelman's disease comes in two forms: unicentric, the most common form, often benign, is manifested by localized lymphadenopathy, very slowly progressive, painless, discovered fortuitously or revealed by a compressive or painful syndrome if it increases in volume , as is the case in our patient.

The multicentric form affects several lymph node sites [9], with systemic manifestations (fever, inflammatory syndrome, hypergammaglobulinemia) especially when associated with Kaposi's sarcoma or malignant non-Hodgkin's lymphoma [10].

The clinico-biological picture of Castelman's disease is non-specific, simulating other pathologies, essentially tumoral, inflammatory or infectious, which must be ruled out first. The radiological aspect also does not suggest the diagnosis and the use of a radioguided biopsy can often make the diagnosis more difficult.

Only surgical excision with an anatomopathological study allows a positive diagnosis to be made; it represents the reference treatment when it allows complete excision of the lesion. In the event that a lymph node is unresectable, due to its size, its location, its adhesions with neighboring structures or close to large vessels, other therapeutic methods can be considered, such as medical treatment with rituximab /steroids or treatment with anti-IL-6 monoclonal antibodies [11]. Radiotherapy remains an alternative in the case of recurrence following incomplete excision [12].

CONCLUSION

Castelman's disease is a heterogeneous group of rare nodal involvement, and is classified as

unicentric or multicentric. The diagnosis seems difficult because of the non-specificity of the clinical and radiological picture. Radical excision remains the first-line treatment of choice in order to allow curative treatment and avoid recurrences.

REFERENCES

1. Castleman, B., Iverson, L., & Menendez, V. P. (1956). Localized mediastinal lymph-node hyperplasia resembling thymoma. *Cancer*, *9*(4), 822-830.
2. Capdevielle, P., Lapprand, M., Derrien, J. P., Courbil, J., Courtois, D., & Barabe, P. (1983). Abdominal forms of Castleman´s disease. *Gastroenterol Clin Biol*, 7(3), 299-306.
3. Akram, W., Degliuomini, J., Wallack, M. K., Huang, S., Okechukwu, E., & Eric, T. (2016). La maladie unicentrique de Castleman se faisant passer pour une tumeur carcinoïde de l'intestin grêle. *Suis Surg*, 82(9), e287-e289.
4. He, X., Wang, Q., Wu, Y., Hu, J., Wang, D., Qi, B., ... & Cheng, J. (2018). Comprehensive analysis of 225 Castleman's diseases in the oral maxillofacial and neck region: a rare disease revisited. *Clinical Oral Investigations*, *22*(3), 1285-1295.
5. Beck, J. T., Hsu, S. M., Wijdenes, J., Bataille, R., Klein, B., Vesole, D., ... & Barlogie, B. (1994). Alleviation of systemic manifestations of Castleman's disease by monoclonal anti-interleukin-6 antibody. *New England Journal of Medicine*, *330*(9), 602-605.
6. Brandt, S. J., Bodine, D. M., Dunbar, C. E., & Nienhuis, A. W. (1990). Dysregulated interleukin 6 expression produces a syndrome resembling Castleman's disease in mice. *The Journal of clinical investigation*, *86*(2), 592-599.
7. Parravinci, C., Corbellino, M., Paulli, M., Magrini, U., Lazzarino, M., Moore, P. S., & Chang, Y. (1997). Expression of a virus-derived cytokine, KSHV vIL-6, in HIV-seronegative Castleman's disease. *The American journal of pathology*, *151*(6), 1517-1522.
8. Soulier, J., Grollet, L., Oksenhendler, E., Cacoub, P., Cazals-Hatem, D., Babinet, P., ... & Sigaux, F. (1995). Kaposi's sarcoma-associated herpesvirus-like DNA sequences in multicentric Castleman's disease [see comments]. *Blood*, 86(4), 1276-1280.
9. Eloueriachi, F., Caidi, M., Zouaidia, F., Ouchen, F., Maidi, M., Fennane, H., ... & Benosman, A. (2012). Castleman disease: unusual location of the chest. *The Pan African Medical Journal*, *12*, 111-111.
10. Lachant, N. A., Sun, N. C., Leong, L. A., Oseas, R. S., & Prince, H. E. (1985). Multicentric angiofollicular lymph node hyperplasia (Castleman's disease) followed by Kaposi's sarcoma in two homosexual males with the acquired immunodeficiency syndrome (AIDS). *American journal of clinical pathology*, *83*(1), 27-33.
11. van Rhee, F., Oksenhendler, E., Srkalovic, G., Voorhees, P., Lim, M., Dispenzieri, A., ... & Fajgenbaum, D. C. (2020). International evidence-based consensus diagnostic and treatment guidelines for unicentric Castleman disease. *Blood advances*, *4*(23), 6039-6050.
12. Beckham, T. H., Yang, J. C., Chau, K. W., Noy, A., & Yahalom, J. (2020). Excellent outcomes with surgery or radiotherapy in the management of Castleman disease including a case of oligocentric disease. *Clinical Lymphoma Myeloma and Leukemia*, *20*(10), 685-689.