A Diagnostic Challenge: Pelvic Castleman's Disease

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Abstract: *Background*: A rare lymph node hyperplasia that resembles malignant tumors is unicentric Castleman's disease. Although, it may appear in any lymph-node basin, the pelvis is an uncommon site for this disease, leading to a challenging preoperative diagnosis.

Case Report: A pelvic tumor was discovered during the infertility sort out of a 31-year-old female. Computed tomography scans showed a pelvic mass with prominent vascular-supply, centrally calcified compressing the urinary bladder. With a malignant pelvic neoplasm diagnosis, a laparotomy was scheduled. At surgery, an 11-cm tumor infiltrating right mayor rectus-abdominis muscle, ipsilateral fallopian tube, and a urinary bladder segment was resected en-bloc. The final histopathology diagnosis was a hyaline-vascular, unicentric Castleman´s disease. After seven years of follow-up, the patient is disease-free.

Conclusion: Unicentric pelvic Castleman's disease resembles malignant tumors and surgery is mandatory for its treatment and diagnosis.

Keywords: Castleman's disease, Pelvis, Surgery, Tumor.

1. INTRODUCTION

As malignant tumors grow they infiltrate surrounding structures. Imaging studies (helical computed tomography scan and magnetic resonance images) may show these invasive features, helping physicians to establish a presumptive preoperative diagnosis. However, there are rare diseases that resemble malignant tumor's behavior, and they are identified in most cases after surgery [1]. This is Castleman's disease case [2].

Castleman's disease (CD) is a rare giant lymph node hyperplasia (giant lymphadenopathy) of unknown origin that macroscopically appears as an encapsulated homogenous mass [2-4].

Microscopically, three subtypes of CD have been identified: the hyaline-vascular, the plasma- cell, and a mixed form. The plasma-cell type has an aggressive course and tends to be multifocal with systemic manifestations. The hyaline-vascular type tends to be localized (unicentric) in one lymph node basin, and it is asymptomatic. Unicentric CD was originally reported in the mediastinum; however, it may happen in any lymph node basin [2-4].

This case will present the diagnostic problems that physician face with this rare disease mimicking a malignant tumor but in a really uncommon anatomical area.

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2. CASE REPORT

A pelvic tumor was founded in a 31-year-old female during infertility sort out investigations. Her medical history included systemic lupus erythematosus, inactive during the last two years. At physical exam, the tumor was in the right lower abdominal quadrant. A pelvic transabdominal ultrasonography detected a heterogeneous tumor with rich vascular-supply and an echogenic central mass, near the uterine fundus. Helical computed tomography scans showed an ovoid pelvic tumor with regular borders, prominent vascular-supply, centrally calcified, compressing the urinary bladder (Figure **1**) associated to right-external iliac lymphadenopathies. With image findings suggesting a malignant tumor, she was referred to surgical oncology. In that service, the image findings were confirmed, and an exploratory laparotomy was scheduled. At surgery, a tumor infiltrating the lower-third of right mayor rectus-abdominis muscle, ipsilateral fallopian tube, and a urinary bladder segment has been revealed. Treatment consisted in tumor en-bloc resection with regional lymphadenectomy (Figure **2**). The histopathology department diagnosis was an unicentric, giant lymphoid hyaline-vascular CD disease (11 cm) invading urinary bladder (Figure **3**). Two years after surgery, with no medical management, she achieved her first full-term pregnancy. Actually, after seven years of follow-up, the patient is disease-free.

3. DISCUSSION

As usually, malignant tumors infiltrate surrounding structures and show rich vascular-supply in image's

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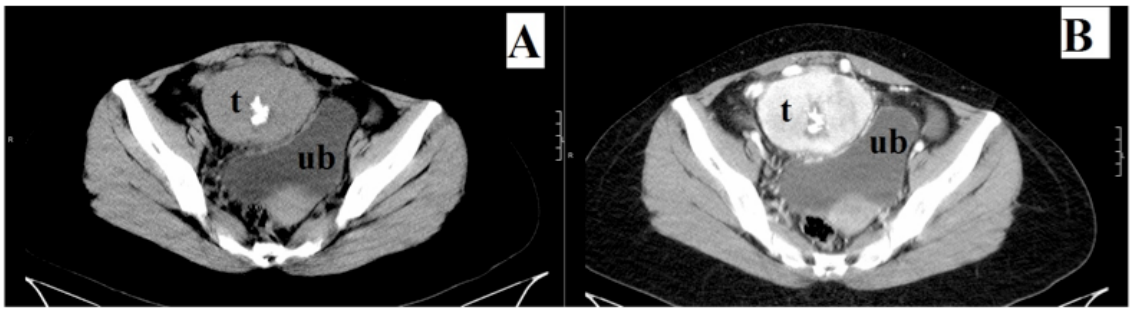

Figure 1: CT image through the pelvis (**A**). Contrast-enhanced image; note tumor´s vascular-supply and central calcification (**B**). "t": tumor. "ub": urinary bladder.

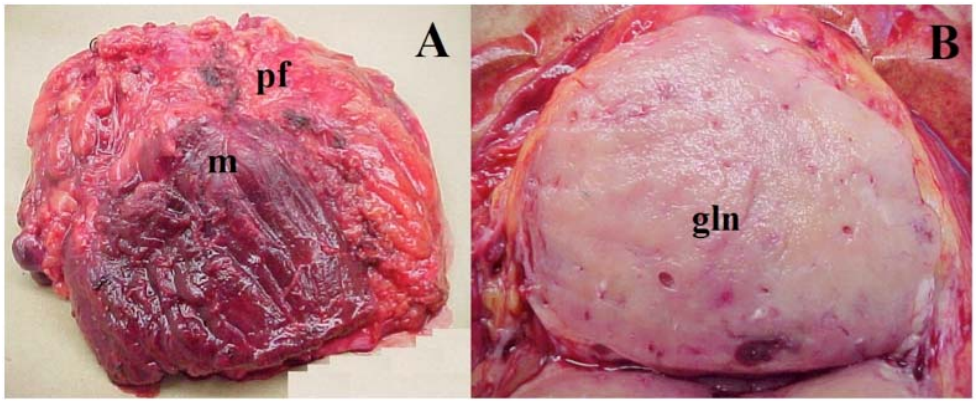

Figure 2: *En-bloc* surgical specimen (**A**). The giant lymph node (**B**). "m": muscle. "pf": pre-peritoneal fat. "gln": giant lymph-node.

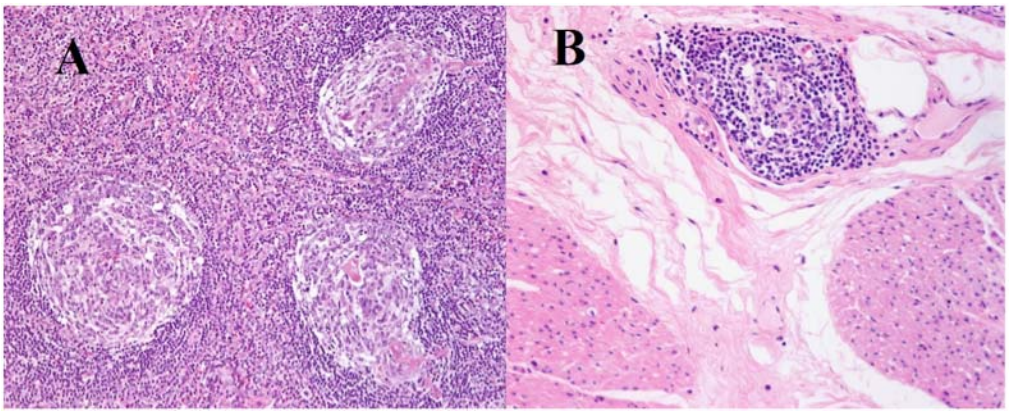

Figure 3: Lymphoid follicles with hyaline material in germinative centers (**A**). Urinary bladder´s wall detail with CD infiltration (**B**). (H-E, 100x).

studies. However, rare diseases may mimic malignant neoplasm's behavior, rendering preoperative diagnosis a challenge.

Clinically, two CD distinct types have been identified: the localized or unicentric and the multicentric. The multicentric form carries a poor prognosis. Instead, unicentric CD involves a single anatomical area and have a better survival prognosis [5]. It generally appears in young healthy individuals with no or few symptoms, Table **1** [6].

Radiographic characteristics of CD are non-specific, but some may help to suspect the diagnosis [7]. Ultrasonography usually shows a hypoechogenic, homogenous mass with well-defined borders and central areas of sharp acoustic shadowing due to calcification. In computed tomography scans, different findings could appear according to lymph node size: when it is ≤5-cm images show a solid, homogenous well delimited mass. However, if it is >5-cm images show a heterogeneous mass with regional lymphadenopathies, central fibrosis or calcifications;

Table 1: Published Pelvic Castleman's Disease Cases

	Author	Age (years)	Gender	Symptoms	Rx findings Tumor size	Treatment	Type	Evolution
1	Takeda 1990	41	♂	NS	Solid mass 4 cm	Resection	PC	Satisfactory
2	McDonald 1996	?	♀	NS	Solid mass 8 cm	Resection	HV	Satisfactory
3	Florez 1997	46	♂	Abdominal pain	Solid mass 5 cm	Resection	HV	Satisfactory
4	Murphy 1997	31	♀	Abdominal pain	Solid mass 7 cm	Resection	HV	Satisfactory
5	Meador 2000	9	♂	Abdominal pain	Solid mas 4.5 cm	Resection	HV	Satisfactory
6	Watson 2000	46	♂	NS	Solid mass 4 cm	Resection	HV	Satisfactory
7	Cammisuli 2003	21	♀	Pelvic pain	Solid mass 4.5 cm	Resection	HV	Satisfactory
8 9	Chang 2004	30 28	♀ ♀	Abdominal pain Vaginal bleeding	Solid mass 3 cm Solid mass 8.4 cm	Resection Resection	HV HV	Satisfactory Satisfactory
10	Hsieh 2004	45	♀	Abdominal pain	Solid mass 6.3 cm	Resection	HV	Satisfactory
11	Nakamura 2004	30	♀	NS	Solid mass 5 cm	Resection	HV	Satisfactory
12	Kakuta 2005	36	♂	Fever	Solid mass 7.5 cm	Resection	PC	Satisfactory
13	Kawamura 2007	57	♂	NS	Solid mass 5 cm	Resection	HV	Satisfactory
14	Zhou 2008	29	♂	NS	Solid mass ? cm	Resection	HV	Satisfactory
15	Hwang 2011	34	♀	Vaginal bleeding	Solid mass 6 cm	Resection	HV	Satisfactory
16	Cascales 2012	19	♀	Pelvic pain	Solid mass 9 cm	Resection	HV	Satisfactory
17	Sato 2013	22	♀	NS	Solid mass 9.5 cm	Resection	HV	Satisfactory
18	Ortiz-Mendoza 2013	39	♀	NS	Solid mass 11 cm	Resection	HV	Satisfactory

HV: hyaline vascular type. NS: no symptoms.

calcifications occurs in approximately 30% of the cases [8,9]. Small or large masses enhance with vascular contrast as a result of hypervascularity [10]. In magnetic resonance imaging, the mass detected is hypo-intense on T1-weighted images and hyper-intense on T2-weighted images, also presenting enhancement with contrast [9,11]. Unfortunately, ultrasonography, computed tomography, and magnetic resonance imaging findings are not pathognomonic, leading to differential radiological diagnosis of a possibly malignant neoplasm, mainly by hypervascularity.

Usually, preoperative work-up such as fine-needle aspiration [12-14] or core-needle biopsies [7] are not helpful to diagnose CD, except for occasional case reports [15]. Even, a surgical specimen could be misdiagnosed as lymphoma in trans-operatory frozen section [16]. Consequently, only definitive histopathology study is the way for a correct diagnosis [14,17].

According to the available literature, pelvic CD is rare and only achieve 2% of all cases [5,18,19]. Talat *et al.* [5] found only five in a review of 404 published cases. Pelvic CD is characterized by tumors >5 cm, with predominance of females and hyaline-vascular subtype, and its diagnosis is only possible after surgery, Table **1** [14,20-33].

The standard therapy of unicentric CD is surgery [5], and it is curative when there is a complete resection [5,34].

To conclude, pelvic CD is a preoperative diagnostic challenge.

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