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Case report

CASE REPORT

Embolization of blood-supply artery followed by surgery for treatment of mesorectal Castleman's disease: case report and literature review

Guanyu Yu†, Fuao Cao†, Haifeng Gong, Peng Liu, Ge Sun and Wei Zhang*

Department of Colorectal Surgery, Changhai Hospital, Shanghai, China

*Corresponding author. Department of Colorectal Surgery, Changhai Hospital, 168 Changhai Road, Yangpu District, Shanghai, China. Tel: +86-13816321041; E-mail: weizhang2000cn@163.com

†These authors contributed equally to this work and are considered co-first authors.

Abstract

A 23-year-old male patient was diagnosed as having a hypervascular pelvic mass by ultrasonography and magnetic resonance examination. A pathology puncture showed vitreous vascular Castleman's disease. Because of concerns about tumor blood supply, embolization under digital subtraction angiography (DSA) was performed on the artery of the pelvic tumor before resection of the mass and surrounding rectum. Castleman's disease of pelvic lymph node (mixed type, mainly hyaline vascular type) was confirmed pathologically from postoperative biopsy. Embolization of the blood-supply artery of a hypervascular mass should be considered before surgery is performed.

Key words: Castleman's disease; mesorectal tumor; embolization

Introduction

Castleman's disease (CD) is a rare disease of unknown etiology and associated with lymph node enlargement [1]. It is generally believed that the disease is associated with human immunodeficiency virus (HIV) and human herpes virus 8 (HHV-8) [2]. In 1954, Castleman reported that the disease occurred in the septum [3]. Further reports confirmed that the most common sites were mediastinal lymph nodes, followed by the neck, axillary and abdominal lymph nodes and some organs outside the nodes such as the throat, vulva and pericardial and intracranial subcutaneous muscle. CD is usually divided into focal and multicenter types clinically and hyaline vascular type, plasma cell type and mixed type pathologically. It is extremely rare that the focal hyaline vascular type of CD occurs within the mesorectum [4].

Case presentation

A 23-year-old male patient with a healthy medical record demonstrated a pelvic mass by ultrasonography during routine examination. During his hospitalization, a large mass which is 4 cm to the anus was found by rectal examination; no abnormal laboratory results were revealed. After the ultrasonic examination of pelvis, a 6.2×6.0 cm hypoechoic and uneven mass was found in the pelvic cavity (Figure 1), and an abnormal signal and a 5.2×6.9 cm hypervascular mass were revealed by magnetic resonance (MR) examination in the rectum and sacrum gap. In

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the MR examination, the mass showed equal signal in T1WI, line-like low signal and separation with central low signal fiber composition in T2WI, and uneven slightly limited signal in diffusion-weighted imaging. In the enhanced image, the mass was strengthened obviously except the central fiber composition. The margin of the lump was clear, with limited surrounding structures, and the rectum was pressed forward (Figure 2). Pathological puncture was performed immediately. An experienced pathologist reported reactive hyperplasia of lymph nodes and vitreous vascular Castleman's disease.

Taking into account the tumor blood supply, embolization under digital subtraction angiography (DSA) was performed on the artery of the pelvic tumor. Angiography of bilateral iliac artery branches showed a round tumor staining shadow. 5 mg dexamethasone, 60 mg epirubicin and a bottle of gelatin sponge (710–1000μm) were injected. The branch of internal iliac arteries blood flow blocking was effective when a second angiography was performed (Figure 3). Resection of anterior rectal and terminal ileal stroma was performed under general anesthesia 7 days after the embolization. After entering the abdominal cavity by a median incision of the lower abdomen, a $4 \times 5 \times 4$ cm^3 medium-texture mass (with numerous nourishing vessels from anterior sacral space in the mesentery of the posterior rectal wall) was found. No enlarged lymph nodes were observed in the surrounding mesentery.

The postoperative gross specimen (Figure 4) showed a tumor situated in the mesentery of the posterior rectal wall 3 cm from the upper and lower resection margins, respectively. The tumor had a diameter of 6 cm with medium, solid and hoar-frost appearance on the surface. Six lymph nodes were found in the mesorectum, with diameters ranging between 0.3 and 1.0 cm. Pathology of the biopsy (Figure 5) confirmed that the mass around the rectum was lymph node with complete structure. Lymphoid follicular hyperplasia of different sizes was seen by microscope, and vitreous blood-vessel fibrous tissue could be seen in the stroma. No obvious change was seen in the rectum. Many vitreous blood vessels were found in the lymph nodes within the mesorectum. Immunohistochemistry results were CD20 (focal +), CD19 (focal +), Bob.1 (focal +), CD3 (partial +), CD5 (partial +), Bcl-2 (partial +), Bcl-6 (+), CD10 (a small amount of focal +), Muml (partial +), CD38 (partial +), CD30 (-), CD34 (vascular +), Ki-67 (+) and PSA (-). According to the clinical symptoms and postoperative pathological and immunohistochemical results, the patient's diagnosis was Castleman's disease of pelvic lymph node (mixed type, mainly hyaline vascular type).

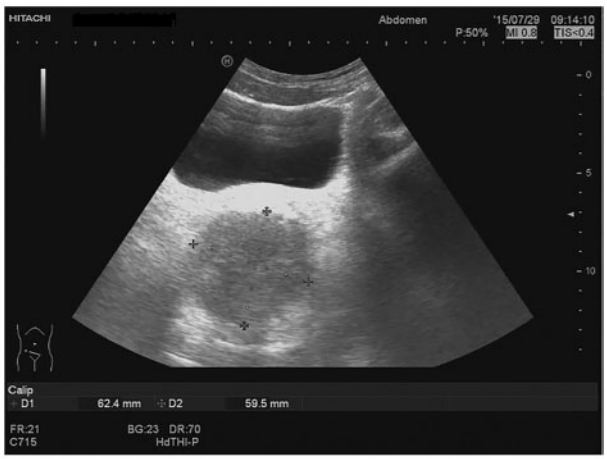

Figure 1. Ultrasonography showed a 6.2 x 6.0 cm uneven and hypoechoic mass in the pelvic cavity

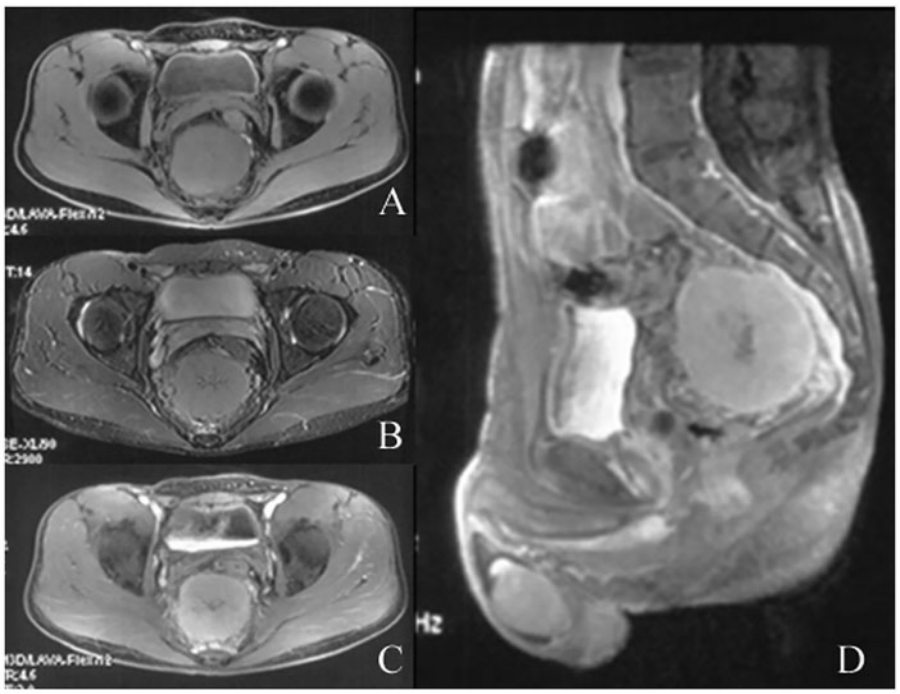

Figure 2. MR examination. (**A**) Horizontal T1WI showed equal signal. (**B**) T2WI showed mixed high signal with line-like low-signal separation. (**C**) Enhanced image: the mass was strengthened obviously except the central fiber composition. (**D**) The margin of the lump is clear with limited surrounding structures, and the rectum is pressed forward

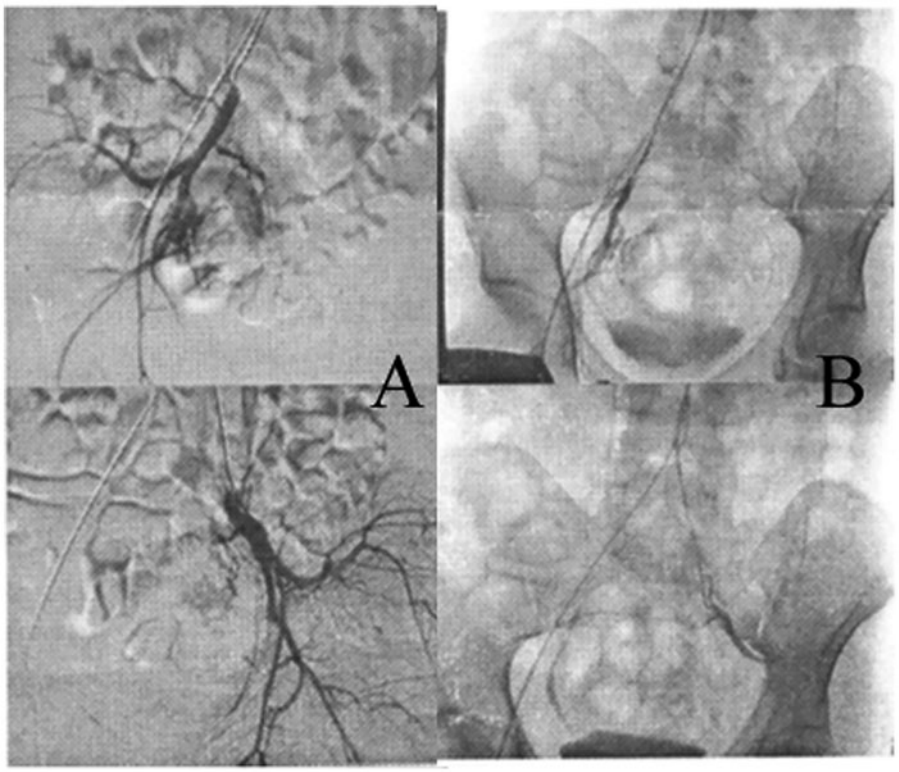

Figure 3. Angiography before and after operation. (**A**) Preoperative angiography showed a round tumor staining shadow supplied by bilateral iliac arteries. (**B**) Blood-flow blocking is good after the operation

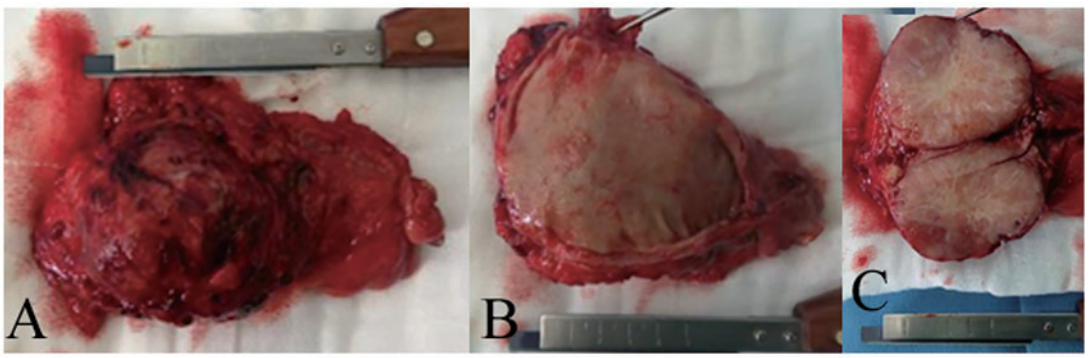

Figure 4. Gross surgical specimen. (**A**) Tumor was in the mesentery of the rectum's posterior wall, 3 cm from the upper and lower resection margin, and its diameter was 6 cm. (**B**) No obvious changes in the rectal lumen. (**C**) A medium, solid and hoar-frost appearance on the surface

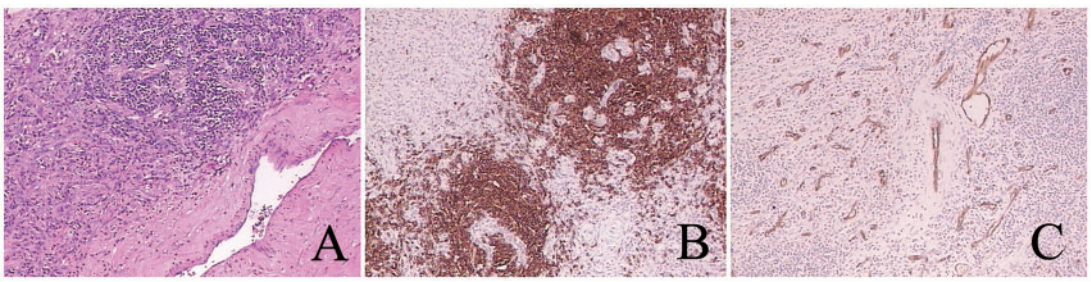

Figure 5. Biopsy pathology. (**A**) The mass around the rectum was lymph node with complete structure. Lymphoid follicular hyperplasia was observed under the microscope with different sizes and vitreous blood vessel fibrous tissue in stroma. (**B**): Immunohistochemistry staining for CD20 (focal +). (**C**) Immunohistochemistry staining for CD34 (vascular +)

Discussion

Although the incidence rate of CD is not officially known, a number of authors have estimated it to be approximately 21 cases per million inhabitants in the USA based on patient cohort results extracted from databases [5,6]. CD can occur in all age groups, but occurs most between ages 10 and 45 years with no difference by sex. There is no consensus on the etiology of CD [7]. Disorders of IL-6 secretion may lead to chronic inflammation and immune deficiency status during HHV-9 or Epstein-Barr virus (EBV) infection, which may cause the disease. Lymphoma and Kaposi's sarcoma may cause autoimmune diseases, which could also be another contributing factor [8]. However, there were no inflammatory or autoimmune problems shown in the patient reported herein.

Pathological nature of CD is complete structure of lymph nodes, lymphoid follicles and vascular proliferation [9]. The most common type of CD is the hyaline-vascular type (90%), which is more common in adults under the age of 35 years. Glass-like follicles and follicular capillary proliferation can be seen under the microscope [1]. The hyaline-vascular type CD mass is mostly focal and is usually found by physical examination (most often accompanied by fatigue and local compression symptoms) [10]. The plasma-cell type is rare and is accompanied by obvious systemic symptoms such as fever, malaise, sweating, weight loss, anemia, thrombocytosis, immunoglobulin and elevated erythrocyte sedimentation rate, etc. The pathological features of this type of CD are dense plasma cells and less vascular stromal cells around the germinal center [1]. The third pathological type is mixed type and includes multiple centers with poor prognosis. Meanwhile CD is also divided clinically into focal and multicenter types. The focal type of CD is easy to resect completely and is followed by better prognosis. The multicentric type is usually found in 50–60 year-old patients, often with hepatosplenomegaly as the first symptom, and has a poor prognosis [1]. The patient reported here had a focal and mixed type (mainly hyaline-vascular type) without the symptoms mentioned above and recovered well after surgery.

In our case, ultrasonography (USG) suggested a hypoechoic mass, whereas MR revealed an equal or low signal in T1WI and high signal in T2WI. Previous reports have suggested that CD can be a linear, tree-like or low-signal mass accompanied by calcification, blood vessels or fiber separation in MR [11–13]. When performing positron emission tomography (PET), typical features in the focal type of CD are low level of fluorodeoxyglucose (FDG) metabolism (which can be identified with significant high-metabolic lesions of malignant tumors), certain infections and inflammatory lesions. The multicentric type of CD show metabolism between low and moderate level, which is difficult to be identified with lymphoma. [14]. However, PET-CT is a good approach for clearly localizing the affected area and identifying the missing lymph nodes. As a result, PET-CT can be helpful for determining clinical classification and guiding treatment in some atypical cases. Our case is in line with the results of USG and MR examination mentioned above. The mass was significantly enhanced, while the enhancement of its central fiber composition was not obvious. Because the tumor's blood supply is very rich, surgeons should take full consideration and operate with delicacy.

The diagnosis of CD mainly relies on pathological biopsy. The multicentric type can be diagnosed by its abnormal symptoms, and the focal type is often diagnosed by physical examination results. Frizzera proposed CD diagnostic criteria: diagnosis of focal type CD includes single-site lymph node enlargement, histopathology with characteristic hyperplasia, no systemic symptoms and anemia, immune globulin protein level elevated (except plasma cell type), long-term survival after resection of tumor and exception of other possible primary diseases. Diagnosis of multicentric-type CD includes characteristic histopathologic changes, significant lymph node (involving multiple peripheral lymph nodes) enlargement, involvement of multiple organs and exception of other possible primary disease [15].

The differential diagnosis of CD mainly includes reaction of lymph node hyperplasia, uterine fibroids, uterine leiomyosarcoma, lymphoma, HIV infection and autoimmune diseases (such as rheumatoid disease or Sjögren's syndrome). The sacral mass can be divided into congenital, inflammatory, neurogenic and bone origin, of which two-thirds are congenital (in congenital cases, two-thirds are progressive, and one-third are neoplastic [16,17]. Mesorectal benign tumors include cyst, lipoma, leiomyoma, teratoma, meningocele, schwannoma and ganglioneuroma. Therefore, immunohistochemical methods can be helpful for an accurate diagnosis [18]. The pathological features of CD are lymphatic follicular hyperplasia, vascular infiltration and follicular plasma cell infiltration.

The treatment of CD includes surgery, radiotherapy, hormonal therapy, immunotherapy and combined chemotherapy. Rituximab, an anti-CD-20 monoclonal antibody, has achieved significant results in treating CD. Cyclophosphamide, vincristine and doxorubicin are commonly used in combined chemotherapy [4,19–21]. Surgical treatment should be considered as the first option in the focal type of CD. If complete resection proves to be difficult, partial resection may be selected. The patients who received focal resection always had better prognosis with low recurrence rate [10,19]. The prognosis of total resection was better in both superficial and deep mass [22]. The results from radiotherapy and chemotherapy are different. The prognosis of patients with malignant type should be followed up for a longer time, which requires further studies [4,19,23].

For surgical treatment of tumors with rich blood supply in the pelvic cavity (especially tumor near the mesentery of the rectum, concerning the complex pelvic structure), surgeons need to be extremely careful and meticulous during the operation. It will greatly affect the prognosis of the patient as well as the process of surgery. Using the example of CD treatment, we suggest that performing preoperative DSA for arterial embolization and blocking the blood supply of the tumor are effective for handling pelvic tumors with rich blood supply. In our case, the preoperative DSA embolization of the internal iliac artery nutrition blood vessels made contribution to the operation process.

Conflict of interest statement: none declared.

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