Castleman disease in the scrotum

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**Abstract**

Castleman disease (CD) is a rare lymphoproliferative disorder that can affect any lymph node in the body, but CD occurring in the scrotum has not been reported to date. We report the case of a 79-year-old man with a painless hard mass in the right scrotum that has been gradually increasing in size for more than 1 year. Abdominopelvic CT scan showed a heterogeneous enhancing mass of 10 cm long in the right scrotum. The patient underwent resection of the right scrotal mass and the pathological diagnosis was Castleman’s disease, plasma cell (PC) type.

1. Introduction

Castleman disease (CD), also known as giant or vascular lobular lymphoid hyperplasia or lymphatic malformation, is a rare non-neoplastic lymphoproliferative disorder of unknown etiology that is easily misdiagnosed. CD occurs most commonly in the mediastinum, cervical regions, and abdominal/pelvic cavity, but can be found in any lymph node station. CD combined with scrotal masses has never been reported so far, and we report a rare case of UCD with plasma cell type with scrotal mass involvement.

2. Case report

A 79-year-old male patient was found to have a painless hard mass in the right scrotum that had been progressively enlarging for more than 1 year without scrotal swelling or pain, and without a history of fever, cough, dyspnea, weight loss, fatigue or night sweats. The patient had a history of gout for more than 2 years and chronic bronchopneumonia for more than 10 years. Physical examination showed an enlarged right scrotum with a palpable mass of about 12 × 6 × 5 cm in size, with clear boundaries, medium texture, movable, no pressure pain, and normal surface skin. Laboratory tests showed lymphocyte count 0.87 × 10^9/L, lymphocyte percentage 16.6%. Other clinical examination and laboratory findings were within normal limit. Abdominopelvic CT showed a mass in the right scrotum measuring approximately 10 × 6 × 6 cm, with heterogeneous enhancement visible on enhancement scans (Fig. 1). In view of the size and the intensive nature of the tumor, The decision was made to surgically remove the lesion.

A tissue mass of approximately 13 × 7 × 6 cm was seen at the time of surgery, including a testicle of approximately 2.5 × 2.5 × 2 cm and a pale white mass of approximately 7 × 6 × 5 cm. Histopathological examination revealed scattered lymphoid follicles in the paratesticular nodes with intervening fibrous tissue hyperplasia and plasma cell hyperplasia (Fig. 2). Immunohistochemistry showed CD138 (plasma cell +), CD38 (plasma cell +), CD3 (perifollicular +), CD20 (follicle +), Lambda (plasma cell scattered +), IgG4 (plasma cell scattered +), IgG (plasma cell +) (IgG4/IgG ratio less than 40%), Ki-67 (about 10% +), MUM1 (plasma cell +), CD20 (follicle +), CD3 (perifollicular +), CD117 (individual +), PLAP (−), CK (−), CD34 (−). Due to the potentially malignant nature of Castleman disease, the patient was advised to seek further treatment in the hematolgy department after surgery (Fig. 3).

3. Discussion

Castleman disease is a heterogeneous nonmalignant lymphoproliferative disorder. The estimated annual incidence of UCD and MCD in the United States is 4300–5200, but other studies have estimated a lower incidence. Males are more susceptible to Multicentric Castleman disease (MCD) than females, but there is no gender preference for Unicentric Castleman disease (UCD). The mean age at diagnosis for patients with UCD (40 years) is usually younger than for patients with MCD (60 years), but all age groups of patients, including young children, can develop any form of CD. The prominent clinical manifestation of the disease is painless lymph node enlargement, with a large diameter of 3–7 cm, or even up to 16 cm. Castleman disease can be divided into UCD and MCD. UCD mostly
presents as isolated enlarged lymph nodes in the mediastinum. Usually either symptoms of compression or incidental finding of nodes are present. MCD presents with Systemic symptoms and multiple lymphadenopathy. Systemic symptoms include recurrent fever, night sweats, anemia, hypergammaglobulinemia, and immunosuppression. Peripheral lymphadenopathy can involve multiple regions of the neck, chest, abdomen, and pelvis.

The diagnosis and treatment of CD have been irregular and misdiagnosed due to the unclear etiology and pathogenesis, as well as the complex clinical presentation and the lack of oncologic markers. CT scans show the location, extent, and number of involved lymph nodes, which are then visualized with uniform high-intensity contrast enhancement. However, it is difficult to differentiate from other diseases and are highly susceptible to misdiagnosis as neurogenic tumors, lymphomas, lymph node metastatic tumors, mesenchymal-derived tumors, thymomas, and gastrointestinal mesenchymal tumors. Magnetic resonance imaging (MRI) can be used to further clarify the soft tissue involvement. Positron emission tomography (PET-CT) can be used not only to obtain a diagnostic sample but also to exclude lymphoma by biopsy of the site with the highest standardized uptake value (SUV). However, postoperative histopathological examination remains the preferred method for final diagnosis.

Surgical resection has been shown to be the gold standard for the treatment of UCD. Complete surgical excision is almost uniformly curative with all symptoms and laboratory abnormalities returning to normal. If surgery is not possible, then irradiation, embolization, or neoadjuvant therapy should be considered. In contrast to UCD, there are many options for the treatment of MCD, but so far there are no official guidelines. Rhee et al. believe that the focus is on sartuximab or tarcizumab (anti-IL-6 monoclonal antibody) or rituximab (anti-CD20 monoclonal antibody) +/- steroids as the main pillar of treatment. Patients with MCD have the possibility of malignancy and can progress to Hodgkin’s lymphoma, non-Hodgkin’s lymphoma or Kaposi’s sarcoma, so the prognosis is poor and long-term follow-up is very important. Related reports indicate that the 10-year survival rate for patients with MCD is only 48.2%, compared with 95% for patients with UCD.

4. Conclusion

Castleman disease can present as a hyperplastic mass in the scrotum, a very unusual localized form that was initially misdiagnosed clinically as a testicular tumor. This case is a plasmocytic type of localized/unicentric lesion in the scrotum that has never been reported before, expanding the differential diagnosis of scrotal masses. Although resection of this lesion appears to be curable, careful follow-up and specialist management is needed to rule out more controversial and aggressive multicentric forms, including the possibility of tumor progression, particularly lymphomatous transformation.

Informed consent form

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Author contributions

Study design: Zhen Song, Zhiyu Zhang, Wenliang Xie, Jun Ouyang. Data acquisition: Zhen Song, Wenliang Xie, Zhang Chen, Jun Ouyang. Drafting of manuscript: Zhen Song, Zhiyu Zhang, Jun Ouyang. Critical revision of the manuscript: Zhen Song, Zhiyu Zhang, Zhang Chen, Jun Ouyang.

Fig. 1. CT of the abdomen revealed a mass in the right scrotum measuring approximately 10 × 6 × 6cm. A: CT plain scan; B: standing CT; C: CT-enhanced arterial phase; D: CT-enhanced venous phase. The density of the lesion: A: 29Hu; C: 37Hu; D: 45Hu.

Fig. 2. Haematoxylin and eosin photomicrograph showed scattered lymphoid follicles, during which fibrous tissue hyperplasia and plasma cell hyperplasia were seen.
Data statement

The data were obtained from the Department of Urology, the Department of Imaging and the Department of Pathology of the First Affiliated Hospital of SU.

Declaration of competing interest

The authors has no conflict of interest.

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Fig. 3. Immunohistochemistry.