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Case Report

Management of Paratesticular Well Differentiated Liposarcoma (Atypical Lipomatous Tumour) without Adjuvant Therapy: Case Report

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Abstract

Less than 200 cases of para-testicular liposarcoma have been reported. Testicular liposarcoma may present with painless or painful intra-scrotal or inguinoscrotal mass of long duration or recent onset but the most common presentation is a painless mass. We present a 58-year-old male patient who presented with the complaint of painless swelling in the right hemiscrotum. Initially, a clinical diagnosis of testicular tumor was made; however, doppler ultrasonography of the scrotum showed paratesticular tumor. Right scrotal orchiectomy was performed and a diagnosis of liposarcoma was made. Histopathological studies confirmed the diagnosis of well-differentiated liposarcoma (atypical lipomatous tumor). No distant metastasis was evident in the whole-body PET-CT scan. Since adjuvant chemotherapy and radiotherapy in the treatment management of the disease is controversial, it was decided to follow up without adjuvant therapy after surgery.

Introduction

Liposarcomas are a heterogeneous group of malignant neoplasms. WHO summarizes five types of liposarcoma, including well-differentiated liposarcoma (well-DLPS) / atypical lipomatous tumor (atypical-LPT), de-differentiated liposarcoma, myxoidliposarcoma, pleomorphic liposarcoma, and myxoid pleomorphic liposarcoma[2]. Well-DLPS is a type of soft tissue sarcoma that arises from adipose tissue. It is characterized by the presence of mature adipocytes and lipoblasts, which resemble primitive fat cells[1]. Well-DLPS and atypical-LPT are synonymous terms describing morphologically and genetically similar malignant neoplasms[2].

The incidence of well-DLPS is relatively low, accounting for approximately 40-45% of all soft tissue sarcomas[2, 6]. Liposarcomas constitute 7% of paratesticular sarcomas and 0.056% of all soft tissue sarcomas[7]. These tumors most commonly occur in the extremities, although other locations may occur elsewhere, such as the retroperitoneum and paratesticular region[2, 8]. The exact cause of well-DLPS is unknown, but certain risk factors such as exposure to radiation and genetic mutations have been identified[9].

These tumors are typically slow-growing and have a low potential for metastasis[3]. The diagnosis of well-DLPS is typically made through imaging studies and biopsy[4]. Surgical resection is the primary treatment for this type of tumor, and adjuvant therapies such as chemotherapy and radiotherapy are not always necessary[5]. The use of adjuvant therapies such as chemotherapy and radiotherapy in the treatment of well-DLPS is controversial. Some studies have suggested that these treatments may not provide significant benefits in terms of recurrence-free survival[10]. However, in cases where surgical resection is not possible or the tumor has a high risk of recurrence, adjuvant therapies may be considered. Overall, the management of well-DLPS requires a multidisciplinary approach, with careful consideration of the individual patient's risk factors and tumor characteristics[11].

Case report

A 58-year-old male patient presented with the complaint of pain in the right hemiscrotum that has been going on for 20 years and has been growing gradually and has been more painful for the last 4 months. With the clinical diagnosis of testicular tumor, the patient was subjected to further examinations.

Radiological and clinical findings

It was reported as a 9 cm right extra-testicular mass in scrotal doppler ultrasonography. Abdominal, pelvis and thorax CT was performed. The clinical and radiological diagnosis was probableliposarcoma, the right testis was normal, which was postero-inferiorly displaced. There was no retroperitoneal or pelvic lymph node enlargement or distant metastasis.

The patient was clinically suspected of paratesticular tumor, and no distant metastases were observed. Blood tests were within normal limits. Orchiectomy, that is, tying and division of the spermatic cord in the deep inguinal ring, and excision of the mass with complete removal of the testis was performed. Surgical findings were a large, firm tumor originating from the paratesticular region, with the right testis and spermatic cord being normal.

Pathological Findings

In the testicular specimen sent to the pathology laboratory, the tumor was macroscopically 10.5 × 6.5 × 5.5 × 5.5 cm in size and located in the paratesticular area, separate from the testicular tissue.It had a yellow-white lipoma-like structure with a well-circumscribed appearance on the cross-sectional surface. Microscopic examination revealed spindle-like, occasionally multinuclear cells, some of which had a very atypical appearance, scattered in a collagenous fibrous stroma. Scattered atypical cells were common in the tumor tissue (Figure 1: A-B).

Immunohistochemical staining showed P16 (+), S100 (-), Vimentin (+) staining in these atypical cells (Figure 1: C). CD34 was positive in some spindle cells. Ki-67 proliferation index was low. There was no lymphovascular invasion. The lesion was well circumscribed but adjacent to the surgical margin in some areas. Testicular tissue and spermatic cord examined together with the lesion had normal histologic margins. When evaluated together with histomorphological findings and immunohistochemical study results, the diagnosis of atypical lipomatous tumor (well differentiated liposarcoma) was reported.

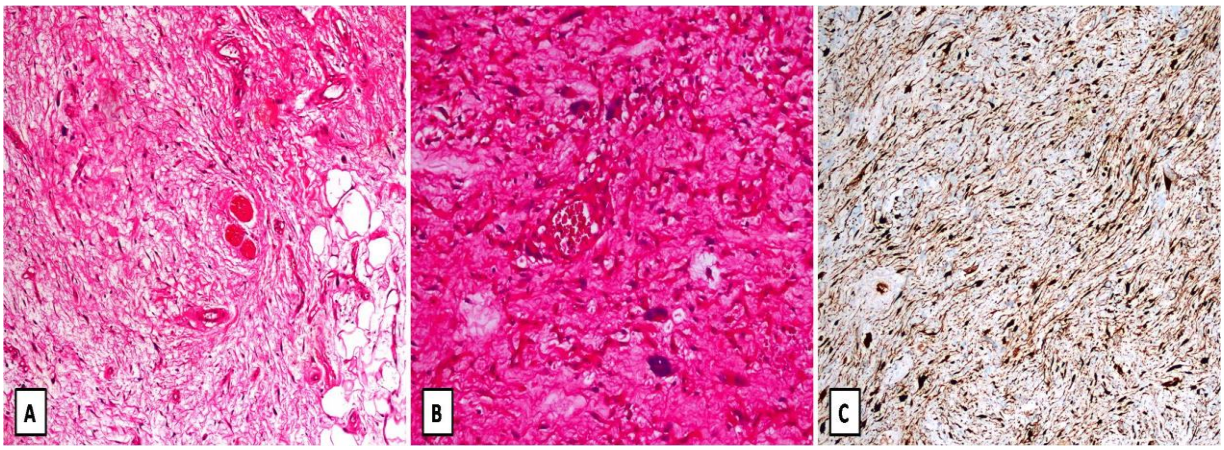

Figure 1: A) Hematoxylin and eosin (H&E) section shows a lipogenic component in the lower right corner of the figure. In other areas, there are atypical cells scattered in the collagenous fibrous stroma (H&Ex100); B) Stromal cells with hyperchromatic nuclei, some with multinuclear atypical appearance (H&Ex200); C) Immunohistochemically P16 positivity in stromal cells (Anti P16 antibody×100).

We did not recommend adjuvant radiotherapy or chemotherapy to the patient, since there was no metastasis in the PET-CT examination performed for restaging in the postoperative follow-ups and there was a pathologically low-grade tumor. The patient was followed up regularly for 9 months after the operation. No local or distant spread has been detected so far.

Discussion

The diagnosis of well-DLPS typically involves imaging tests, such as computed tomography (CT) scans or magnetic resonance imaging (MRI), to identify the location and extent of the tumor. A biopsy is then performed to confirm the diagnosis. Histopathological examination of the biopsy sample is used to determine the subtype of liposarcoma and its grade, which is based on the degree of differentiation and the presence of atypical cells[(2)]. Staging is also determined based on the size and location of the tumor, as well as the presence of metastases.

Treatment forwell-DLPS typically involves surgical resection of the tumor, with the goal of achieving negative margins[(2)]. Adjuvant treatment, such as radiotherapy or chemotherapy, may be considered in certain cases, such as for high-grade tumors or those with a high risk of local recurrence[(12)]. However, the effectiveness of adjuvant treatment for

well-DLPS is controversial, with some studies suggesting that standard chemotherapy regimens are generally ineffective[12].

The prognosis for patients with well-DLPS is generally favorable, with a low risk of metastasis. However, the location of the tumor can impact treatment options and outcomes. For example, paratesticular and spermatic cord tumors may be misdiagnosed preoperatively due to their clinical presentations, leading to delays in treatment[13]. Overall, a multidisciplinary approach to treatment, including surgery and adjuvant therapy as appropriate, is necessary to achieve the best possible outcomes for patients with well-differentiated liposarcoma.

Well-DLPS of the testis is primarily managed with surgical intervention. Surgery is considered the first-line treatment, and the mainstay of treatment is often surgical resection, combined with histology-specific chemotherapy and radiotherapy, if necessary[12]. The surgical technique typically involves wide excision and complete removal of the testes and spermatic cord[14]. However, extended resection carries a higher risk of complications and should be considered on a case-by-case basis. Some reports have shown that radiation therapy and chemotherapy may be useful in certain cases, but surgery remains the primary treatment option, and there is no beneficial role for adjuvant chemotherapy or radiation after resection[12, 15-20].

The surgical approach may vary depending on the location, size and extent of the tumor.

Radiotherapy can be an effective treatment option for well-differentiated liposarcoma, particularly when used in combination with surgery. Indications for radiotherapy in the treatment of well-DLPS include the presence of residual disease following surgery, high-grade tumors, and tumors that are difficult to resect completely. Adjuvant radiotherapy can also be used to reduce the risk of locoregional recurrence in cases where complete surgical removal of the tumor is not possible. There are two main types of radiotherapy used in the treatment of well-differentiated liposarcoma: external beam radiation therapy (EBRT) and brachytherapy. EBRT involves directing high-energy radiation beams at the tumor from outside the body, while brachytherapy involves placing radioactive sources directly into the tumor or surrounding tissue. Both types of radiotherapy can cause side effects, including fatigue, skin irritation, and damage to surrounding tissues.

Radiotherapy has generally been used for local control in liposarcoma. Liposarcomas are the most radiation sensitive sarcomas and some cases have achieved remission with

radiotherapy alone, but the results in para-testicular liposarcoma are less clear. Radiation therapy is thought to be more aggressive behavior in addition to surgery; may be recommended in cases such as residual disease, inadequate or positive surgical margins, recurrent disease, high grade or lymphatic invasion[16-18].

In general, radiotherapy is not recommended as the primary treatment option for well-differentiated testicular liposarcoma. This is because of the low response rate of these tumors to radiotherapy. While preoperative radiotherapy can reduce local recurrence rates, it does not improve overall survival[19]. Adjuvant radiotherapy is only recommended for large, high-grade sarcomas of the extremity[20]. A critical narrative review of radiotherapy for retroperitoneal soft tissue sarcoma also found limited evidence supporting the use of radiotherapy for this type of cancer[15].

Several previous studies have reported that adjuvant radiotherapy is effective after surgical treatment for disease control[21-24]. However, the efficacy of adjuvant radiotherapy for paratesticular liposarcoma remains unclear[12, 21, 25-27].

The effects of adjuvant radiation therapy after surgery remain unclear. Even in the subgroup analysis of patients with positive surgical margins in the largest meta-analysis of paratesticular liposarcoma, adjuvant radiation therapy had no statistically significant effect on relapse-free survival[12].

Chemotherapy may be indicated for patients with advanced or metastatic disease, or for those who are not able to undergo surgery. However, there is no consensus on the use of chemotherapy for well-differentiated liposarcoma, and its effectiveness remains unclear[17, 28].Chemotherapy for well-DLPS may involve the use of various drugs, such as doxorubicin and ifosfamide, either alone or in combination[28]. The choice of chemotherapy regimen depends on several factors, including the patient's age, overall health, and the extent of the cancer. Overall, the use of chemotherapy for well-DLPS remains controversial, and its effectiveness in improving patient outcomes is still being studied. While chemotherapy may be indicated for certain patients, such as those with advanced disease, it is not considered a standard treatment option for those with localized well-differentiated liposarcoma. Surgical removal of the tumor, with or without adjuvant radiotherapy, remains the primary treatment modality for well-differentiated liposarcoma.

Hereby, surgical resection with negative resection margins is the only treatment modality that provides a chance of cure, while chemotherapy or radiation therapy is often

used as adjuvant therapy in cases with poor prognostic factors. In addition adjuvant radiotherapy is recommended in cases with multiple local recurrences, positive margins, and/or poor prognostic factors.

There is no definitive role for other treatment modalities such as chemotherapy, retroperitoneal lymph node dissection (RPLND), and radiotherapy in the management of well-DLPS[29].

Follow-up and surveillance are essential components of the management of well-DLPS (30). A follow-up period of up to a decade after surgery is recommended[2, 30]. During this time, imaging studies such as CT and MRImay be used to monitor for recurrence and metastasis. In cases of retroperitoneal sarcoma, surveillance imaging is recommended every 3-6 months for the first 2 years, every 6-12 months for the next 3 years, and annually thereafter [30]. Thus, regular follow-up and surveillance are important for detecting recurrence and metastasis early and providing prompt treatment.

Conclusion

Complete surgical resection remains the definitive treatment for resectable,localized and well differentiated liposarcomas. Adjuvant radiotherapy / chemotherapy may not be required for the management of this particular tumor type, which requires a multidisciplinary approach. Use of adjuvant therapyshould be made on a case-by-case basis, taking into account the individual patient's medical history, tumor characteristics, and other factors. Further research is needed to fully understand the role of radiotherapy and chemotherapy in treating well-DLPS and to develop more effective treatment strategies for this rare and challenging disease.

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