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Paratesticular Rhabdomyosarcoma in an Adult Patient: Case Report and Literature Review

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***Abstract*—Intrascrotal rhabdomyosarcoma in adults is a rare tumor with high aggression and a poor prognosis. The diagnosis is made by the anatomopathological study. The treatment must be multimodal and involves surgery, chemotherapy and radiotherapy. We report our patient's case and review the relevant literature to improve the understanding of this rare disease. Here we describe the case of a 22-year-old man with a solid right paratesticular mass. He underwent right sided orchiectomy with histopathology revealing paratesticuler embryonal rhabdomyosarcoma. Postoperative staging work up revealed multiple retroperitoneal lymph nodes and lung metastasis. Palliative chemotherapy was performed.**

***Index Terms*—Rhabdomyosarcoma, paratesticular, orchiectomy, chemotherapy.**

I. INTRODUCTION

Rhabdomyosarcoma (RMS) is the most common soft tissue tumor in children, but it is rare in adults [1, 2]. Intrascrotal tumors originate primarily from germ cells, whereas non-germinal cell tumors are uncommon [3]. Paratesticular RMS in an adult patient is an extremely rare condition, with only few reported cases in literature [3]. It is an intrascrotal tumor that is localized in paratesticular structures such as the epididymis or spermatic cord. These tumors are characterized by their malignant potential with rapid spread, and requiring early diagnosis and treatment, especially in the aged patients, where prognosis is worse [4]. The treatment must be multimodal and involves surgery, chemotherapy and radiotherapy.

The aim of presenting this case report is to highlight clinical presentation and management of paratesticular RMS in adult patient.

II. CASE REPORT

A 22-year-old young man with no significant previous medical history, presented to urologist and complained that he had a painless right scrotal mass evolved over 4 months.

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The clinical examination revealed enlarged right scrotum with a hard, tender mass adhering to the right testis and epididymis, painless on palpation and without local inflammatory signs. The trans-illumination is negative, the bilateral inguinal lymph nodes were unpalpable and no significant findings were detected on the other physical examinations.

The ultrasound performed shows right intrascrotal tissular mass of heterogeneous structure, echoic, containing small areas of necrosis, with small vascularity especially at the periphery. Pelvic computed tomography (CT) revealed right intra scrotal tumor process, well limited, of tissue density, which is enhanced heterogeneously by delimiting some areas of necrosis after injection of contrast product (figure 1).

Right radical orchiectomy by inguinal approach was performed. A histological examination of the lesion showed round- to- oval cells with mild nuclear atypia, scant amount of cytoplasm, and with areas of necrosis. Immunohistochemistry showed the tumor tissue to be negative for CD117, pancytokeratin, and placental alkaline phosphatase. Positive immunohistochemistry results were found for CD56, myogenin, myoblast determination protein 1 (MyoD1), desmin, and Ki-67 (70%+). The histopathologic diagnosis was embryonal RMS.

Our patient's levels of tumoral markers such as alpha- fetoprotein, beta- human chorionic gonadotropin and serum lactate dehydrogenase was normal.

The Postoperative staging work up revealed multiple enlarged lobulated retroperitoneal lymph nodes, the largest observed around the left paraaortic recess measuring 5.2 cm, and lung metastasis.

The patient received chemotherapy with VAC regimen (Vincristine, Adriamycin, and Cyclophosphamide) in combination with haematopoietic growth factor. Each chemotherapy session was conducted for every 3 weeks. after completing the 6 cycles of chemotherapy, the patient presented ganglionic progression and refused further treatment.

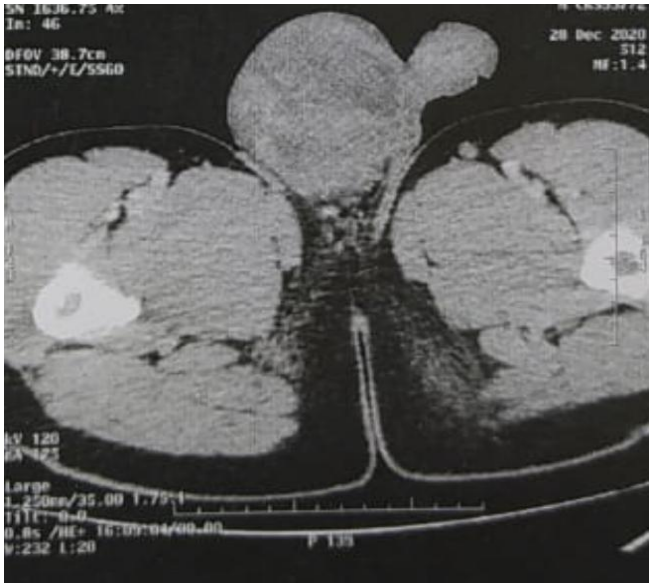

Figure 1: Computed tomographic scan (CT) showing right intra scrotal tumor process

III. DISCUSSION

Rhabdomyosarcoma (RMS) is one of the most common pediatric tumors, comprising up to half of all soft tissue sarcomas [1, 2]. There are two frequency peaks found for the development of RMS, the first at the age of 4 years and the second at the age of 18 years. There is no predilection for race [5]. However, adult RMS is relatively rare, accounting for only 3% of all soft tissue sarcomas [1, 2].

Paratesticular RMS accounts for about 7% of genitourinary RMS [6]. It is thought to arise from the mesenchymal elements of the epididymis or spermatic cord [7]. According to the international classification of RMS, the most common histologic types of RMS are botryoid embryonal, embryonal, spindle cell embryonal, anaplastic, and alveolar [8]. Primitif paratesticular localization is considered to be of good prognosis compared to other rhabdomyosarcomas, despite the frequency of retroperitoneal lymph node involvement. Its superficial localization allows a rapid diagnosis and consequently an often-complete resection of the tumor. Local spread is very early and distant spread is lymphatic and bloodstream. The most frequent metastatic sites are retroperitoneal lymph nodes, lungs, liver and bones [9-11].

The typical clinical manifestation for paratesticular RMS is a painless epididymal mass, or nonspecific symptoms, such as decreased appetite, fatigue, inguinal lymphadenopathy, and weight loss. A paratesticular RMS can cause pain when it oppresses the nerve. However, pain is extremely uncommon and present in only 7% of the cases, whereas a hydrocele may be also a rare presentation [12-14]. When paratesticular RMS present with painful unilateral scrotal swelling symptoms, it often leads to a misdiagnosis of epididymitis.

The determination of tumor markers (HCG, LDH, FP) must be part of the assessment of any testicular tumor, but it is often normal in paratesticular tumors. There are no tumor markers that can help approch the diagnosis, which is based only on the histological examination of the orchiectomy [10].

Scrotal sonography is the initial imaging modality for the evaluation of a scrotal mass. This imaging modality shows a mass with heterogeneous echogenicity and inguinoscrotal extension in 80% of the cases [15]. Wood and Dewbury reported a case in which ultrasonography revealed increased epididymal and testicular blood flow, consistent with epididymo-orchitis [16]. Further evaluation is very important in patients presenting with epididymitis symptom, especially when patients not respond well with antibiotic therapy.

The CT scan, MRI, or PET/CT is regularly used to evaluate distant metastases [17,18]. Both CT and MRI can be used to assess the site, dimensions, and any distant metastases of the tumor [19]. PET/CT can give accurate information about distant metastases of the malignancy. However, none of them is a confirmatory method.

The limitations of traditional diagnostic methods make accurate presurgical diagnosis of paratesticular RMS difficult, and definitive diagnoses therefore depend on postoperative pathologic examination. Observation of the gross specimen is the key to confirming the origin of the tumor [20]. At the opposite of other localizations, the biopsy of paratesticular RMS is prohibited because of the high risk of dissemination [21]. There are four histological types of rhabdomyosarcoma: pleomorphic, alveolar, botryoidal, and embryonal. The characteristic cell is rhabdomyoblast, which is not necessary for the diagnosis.

Whenever rhabdomyoblasts are not present, immunohistochemical investigations are conducted using a panel of antibodies [22]. Immunohistochemical staining is now the optimal diagnostic method for RMS, which is typically positive for one or more muscle-specific markers, including desmin, muscle-specific actin, MyoD1, myoglobin, and/ or myogenin [7,23,24]. These markers are crucial for the differential diagnosis of RMS from other primary mesenchymal and germ cell tumors also exhibiting rhabdomyoblastic differentiation [25].

Intrascrotal RMS has a poor prognosis. The 1-year overall survival (OS) rate is 68%, and the 5-year OS rate is 30% [3]. Retroperitoneal lymph node (RPLN) metastasis is an important prognostic factor. Ferrari et al. found that the 5-year disease free survival was 97% for patients without RPLN metastases and 42% for those with metastasis [26]. Another prognostic factor is the age of the patient. Compared with pediatric patients with RMS, adults with RMS of any organ have significantly worse long-term outcomes [7].

According to the Intergroup Rhabdomyosarcoma Study Group (IRSG), surgicopathologic staging of RMS is predictive of outcome [27] which also could guide the treatment. Some data indicate that staining for myogenin correlate with decreased survival [25]. Literature also reported that anatomic site was also a significant prognostic indicator [28]. Currently, primary paratesticular RMS generally have a better prognosis and a higher survival rate than other RMS [29].

Because RMS rarely occurs in adults, treatments used in pediatric patients are often applied to adult cases. The Intergroup Rhabdomyosarcoma Study Group (IRSG) classifies RMS into four groups by their pathologic margins

and lymph node metastasis status, and it recommends respective targeted treatments [30].

From the data of the literature, the standard treatment for intrascrotal RMS is radical orchiectomy combined with adjuvant chemotherapy and radiotherapy [31].

The role of retro peritoneal lymph node dissection (RPLND) is controversial. Some like Ferrari et al. consider that it is not necessary because of the low rate of retro peritoneal lymph node invasion as well as the demonstrated role of concomitant chemotherapy in eradicating micro metastatic disease [32].

Patients with unresectable tumors who undergo treatment with chemotherapy should be considered for surgery after downgrading.

In the metastatic setting, many protocols of chemotherapy have been tried. VAC, IVA, and VIE protocols (V: vincristine, A: actinomycin, I: ifosfamide, E: etoposide, and C: cyclophosphamide) and better results were observed with VAC protocol [33-35].

Radiotherapy is recommended more commonly to control local recurrence, metastasis, or for unfavorable histology, such as alveolar rhabdomyosarcoma [36].

The combination of all three modalities, that is complete surgical resection, chemotherapy and radiotherapy have greatly enhanced the survival rate in para-testicular RMS with no significant long-term complications.

IV. Conclusion

Para-testicular RMS is a rare and particularly aggressive disease, manifesting primarily in children and adolescents but can rarely affect adults as well. When localized, this disease has a good prognosis and where metastatic, it has a very dismal outcome. The definitive diagnosis of RMS depends on postoperative pathology, physical examination and imaging tests can establish clinical suspicion and detect metastases. Its therapeutic management is not yet well established and is currently based on that of the child. Systemic chemotherapy is essential in both early and advanced disease and has resulted in improved survival outcomes.

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