Spermatic cord tumors — review of the literature

ABSTRACT

In this article, we discuss benign and malignant spermatic cord tumors. We attempted to compile this rare group of diseases by reviewing the international literature. Tumors of the spermatic cord are found very rarely. However, it is important to be aware of their occurrence, as they can cause a protrusion in the inguinal area. They are usually misdiagnosed as an inguinal hernia. The most common tumors in this area are benign — usually they are lipomas. In 20–70% of cases, adipose tumors accompany an inguinal hernia. Therefore, they should be kept in mind whenever a patient presents with symptoms of herniation in the inguinal region. Tumors of the spermatic cord may also involve the scrotum and manifest themselves as testicular hydrocele. Such a tumor is, for example, aggressive angiomyxoma. It is a locally malignant tumor that tends to infiltrate and compress the surrounding tissues but does not tend to give metastasis, therefore according to the WHO classification it is a benign tumor. However, malignant tumors such as rhabdomyosarcoma, which is the most common malignant neoplasm of testicular appendages, can also be located in the spermatic cord. The second most common soft tissue sarcoma is leiomyosarcoma, with poor initial prognosis, or metastases of malignant tumors from other organs, e.g. renal adenocarcinoma. As the prognosis for malignant tumors of the spermatic cord is generally dependent on the stage at the time of diagnosis, oncological vigilance and early diagnosis allow for faster detection of these tumors, which may improve the prognosis of patients with tumors in this location.

Key words: benign tumors, malignant tumors, tumor, spermatic cord, vas deferens

Introduction

Tumors of the spermatic cord are a rare heterogeneous group of diseases. They are usually benign lesions. However, in the minority of cases, malignant neoplasms and metastases of the neoplastic process from another location, such as metastases of renal adenocarcinoma, may also occur within the spermatic cord [1].

Non-malignant tumors of the spermatic cord include lipoma, leiomyoma, rhabdomyoma, cellular angiofibroma, haemangioma, and aggressive angiomyxoma [2].

Malignant tumors of the spermatic cord are very rarely described in the literature.

Malignant lesions originating from adipose tissue and involving the spermatic cord are well-differentiated liposarcoma, dedifferentiated liposarcoma, myxoid liposarcoma, and pleomorphic liposarcoma. A very rare tumor of the spermatic cord originating from smooth muscle tissue that can also spread to the spermatic cord is leiomyosarcoma [2, 3].

The spermatic cord may also be affected by tumors such as rhabdomyosarcoma, desmoplastic small round cell tumor, and metastatic tumors [2, 3].
Benign tumors of the spermatic cord

Lipoma

Lipomas of the spermatic cord are the most common benign tumors in the inguinal canal [4–6]. The incidence of spermatic cord lipoma (without a hernia sac) is 1–8% [7]. A spermatic cord lipoma usually accompanies an inguinal hernia (20–70% of cases with or without a hernia sac) [8–10]. These tumors may increase the size of the inguinal hernia and be misdiagnosed as just an inguinal hernia [8–10]. They are often diagnosed during hernioplasty [8]. A spermatic cord lipoma is usually located deep in the testicular levator muscle and fascia [6].

This tumor is a pre-peritoneal adipose tissue that shows communication with the spermatic cord. This fat merges with the fatty layer within the internal seminal fascia [11, 12]. This is not a true lipoma, which is a benign tumor of adipocytes confined to the inguinal canal and showing no connection to the retroperitoneal fat [11]. However, the term “lipoma spermatic cord” has become entrenched in clinical terminology and is still used. The term “true” adiposity can only be applied to adipomas that are confined to the inguinal canal and show no continuity with pre- or retroperitoneal fat [4–6].

Spermatic cord lipomas usually cause typical symptoms like an inguinal hernia i.e. bulging and pain, so it is recommended that they be treated in the same way as an inguinal hernia [8, 13]. They should also always be clinically suspected when patients report groin pain in the absence of a bulge in the inguinal region [8].

Ultrasoundography (USG) is usually able to detect spermatic cord lipoma. A lipoma on ultrasound is visible as a hyperechoic mass [14]. In doubtful situations, computed tomography (CT) or magnetic resonance imaging (MRI) are recommended [15, 16].

Leiomyoma

Leiomyoma is a benign smooth muscle tumor. This tumor can occur in almost any organ but is most commonly described in the uterus [17, 18]. A review of the literature has so far described single cases of leiomyoma in the genitourinary system — usually in the bladder, epididymis, prostate, testis, and penis [19–24]. Leiomyoma of the spermatic cord is very rare. Since 1949, only three cases of this tumor in the spermatic cord have been described in the literature.

Based on the cases described, it usually manifests as a protrusion mimicking an inguinal hernia with or without scrotal involvement [25]. The authors of these articles recommend that the treatment of leiomyomas should be individual, but once the lesion has been resected, treatment such as that of an inguinal hernia is recommended [25].

Rhabdomyoma

A rhabdomyoma is a benign tumor that accounts for only 2% of all tumors arising from striated muscle [1]. This tumor very rarely affects the spermatic cord. To date, two cases of rhabdomyoma of the spermatic cord have been described [26, 27]. One of these cases was a 67-year-old man with an adult-type subtype. In this patient, the tumor originated from the testicular levator muscle. Seven years later, a case study on a 28-year-old man with rhabdomyoma of the spermatic cord was published [27], and so far no further cases have been described.

In general, rhabdomyomas are slowly growing tumors with a low tendency to recur after radical excision [26].

Cellular angiofibroma

Cellular angiofibroma in men can occur in the spermatic cord (vas deferens) but can also involve the epididymis, the vaginal sheath, and the inguinal region and scrotum [28]. It is a benign mesenchymal tumor that lacks differentiation into smooth muscle, nerves, epithelium, and myoepithelium [29, 30].

It is usually characterized by a benign course with slow growth and no tendency to metastasize [31]. However, this tumor may recur. It is often misdiagnosed as an inguinal hernia [32].

For the treatment of cellular angiofibroma of the spermatic cord and if the tumor involves other perinuclear structures during resection of the lesion, testis-sparing methods are recommended [32].

Hemangioma

Hemangioma is a very rare benign tumor of the spermatic cord. It is characterized by slow growth. [33]. This tumor is usually localized in the inguinal region and scrotum [34]. Since 2009, several cases of anastomosing hemangioma of the spermatic cord have been described [35]. This is a rare subtype of spermatic cord hemangioma. Usually, this tumor is localized in the kidney [36–38]. It is a tumor that is characterized by a benign course [35, 39, 40], but it tends to recur. Nevertheless, isolated cases of anastomosing hemangioma and metastasis have been described in the literature. This tumor presents diagnostic difficulties as it shares many features with malignant sarcomas [41].

Symptoms of spermatic cord hemangioma are usually pain at the tumor site, a palpable protrusion, and sometimes hematuria is also found. On ultrasound diagnosis, anastomosing hemangioma is usually a hypoechogenic or anechoic cystic lesion [36, 37]. It may also show increased marginal asciarization with the use of Doppler techniques [38]. In contrast, on CT anastomo-
Malignant tumors of the spermatic cord

Liposarcoma

Liposarcoma is the most common soft tissue sarcoma. This tumor accounts for 9.8–18% of soft tissue sarcomas. The peak incidence of this tumor is between 40 and 60 years of age. The incidence of this tumor in the scrotum is 3.6%, while in the spermatic cord, the tumor is located with a frequency of 76%. Other locations include the testicular membrane (20%) and epididymis (4%) [46].

According to the World Health Organisation (WHO) classification [2], there are several subtypes of adenosarcoma: well-differentiated liposarcoma, dedifferentiated liposarcoma, myxoid liposarcoma, pleomorphic liposarcoma.

A symptom of liposarcoma may be a painless tumor of the inguinal or scrotal region. On physical examination, a hard, non-painful tumor is palpable [46]. A well-differentiated liposarcoma requires confirmation of MDM2 amplification during diagnosis [47]. Dedifferentiated liposarcoma and well-differentiated liposarcoma share amplifications in the chromosomal region 12q13-15. These amplifications involve MDM2 (100%) and often CDK4 (90%). These amplifications can be detected by fluorescence in situ hybridization (FISH), which is now recognized as the standard for differential diagnosis [48, 49]. However, FISH requires specific equipment that is only available in specialized centers. Therefore, immunohistochemistry (IHC) can be used as an easier method in application and availability [50].

In general, the treatment of liposarcoma depends on the stage of the tumor and histological type. As is well known, soft tissue sarcomas have a high tendency to recur even after previous resection [3]. Generally, surgical resection of the tumor is the primary method of treatment, but this is not always possible and sometimes, even if performed, is insufficient due to local recurrence [51]. Usually, the efficacy of adjuvant chemotherapy (CTH) and radiotherapy (RTH) is low. Single reports have described recurrences more than 10 years after tumor resection, so long-term careful follow-up of the patient after treatment is required [46].

The prognosis in liposarcoma for all sites is dependent on tumor histology. Well-differentiated liposarcoma and myxoid liposarcoma have a better prognosis than other histological subtypes of this tumor.

According to the literature, 5-year survival in well-differentiated liposarcoma is approximately 85%, in the myxoid subtype it is 77% and in other subtypes of this tumor, it is 20% [52].

However, liposarcomas can undergo differentiation. They may most commonly differentiate approximately 7.7 years after the diagnosis of the well-differentiated type. When a liposarcoma differentiates, 5-year survival drops to 28%. Differentiation occurs most commonly in recurrent tumor metastases [53, 54].

Leiomyosarcoma

Leiomyosarcoma of the spermatic cord is a rare malignant tumor of this region. However, a review of the literature suggests that 75% of soft tissue sarcomas in men originate from the spermatic cord [5]. Usually, leiomyosarcoma of the spermatic cord originates from its distal segment.

Leiomyosarcoma of the spermatic cord may imitate an incarcerated inguinal hernia. In addition, it can be confused with an epididymal cyst, a lipoma spermatic cord, and epididymo-orchitis. The clinical presentation of this neoplasm is usually vague and atypical. The patient may report the presence of a palpable
painless mass in the groin and scrotal region [55]. The peak incidence is between 50 and 60 years of age [56]. Leiomyosarcoma of the spermatic cord is the second most common soft tissue sarcoma. To date, 113 cases of leiomyosarcoma of the spermatic cord have been described worldwide [57].

The neoplasm spreads in three ways, through local-regional, hematogenous, and lymphatic routes. Local spread is the most common. Hematogenous spread generally involves the liver and lungs while lymphatic spread involves the external iliac, subcostal, paraaortic and common iliac nodes [58]. Ultrasound and CT are helpful in the diagnosis, but the final diagnosis is based on histopathology and immunohistochemistry [55].

Due to its rarity, there is no official position on how to treat this tumor [56]. Orchidectomy with excision of the spermatic cord up to the deep inguinal ring is recommended when the tumor is resectable [57]. Local recurrence is estimated to occur in 30–50% of cases [59]. In the literature, recurrence of this tumor has been described even 15 years after the initial diagnosis [60].

There is also no official position as to the use of RTH. Some authors advocate the use of adjuvant radiotherapy after orchidectomy to reduce local recurrence [61].

Chemotherapy is often used when metastases are present [56]. In contrast, lymphadenectomy of the surrounding lymph nodes is only recommended if they are enlarged. The overall prognosis in leiomyosarcoma, at any location, is poor.

Rhabdomyosarcoma

Rhabdomyosarcoma is a rare malignant tumor that can also occur in the spermatic cord. To date, 62 cases of this neoplasm in the spermatic cord have been described [62, 63]. Rhabdomyosarcoma is the most common malignant neoplasm of the testicular appendages in patients aged 7 to 36 years, and its peak incidence is in the first two decades of life [64, 65]. The WHO classification of tumors of the spermatic cord and testicular appendages distinguishes four subtypes of rhabdomyosarcoma [2]: embryonal type, alveolar type, spindle cell type, and pleomorphic type.

Embryonal type

The embryonal type of rhabdomyosarcoma is the most common subtype of this tumor in children and young adults [63].

Pleomorphic type

In adults, it is most commonly localized in the deep tissues of the extremities [3].

Alveolar type

In this type of rhabdomyosarcoma, chromosomal translocations are most commonly found. The usual translocation found is t(2;13) (q35;q14) with the formation of a fusion between the PAX3 gene on chromosome 2 and the FKHR gene found on chromosome 13. Another translocation found is t(1:13) (p36;q14).

PAX3 acts as a cascade gene for other genes controlling differentiation into skeletal muscle. Tumor development most likely occurs as a result of disruption of the muscle differentiation process by a chimeric protein formed after PAX3-FKHR fusion [66, 67].

Spindle cell type

In children, it is most commonly localized within the scrotal sac [68]. Rhabdomyosarcoma usually manifests as a painless palpable tumor in the epididymal region or as an enlargement of the scrotum on the tumor side [63]. In addition, enlarged inguinal lymph nodes are often found on physical examination also on the side of the lesion.

In the treatment of rhabdomyosarcoma of the spermatic cord, there are no strict management guidelines due to the rarity of this tumor. Most authors advocate the need for orchidectomy regardless of tumor stage. Lymph node metastases may affect up to 50% of patients with this tumor, so lymphadenectomy of the involved retroperitoneal lymph nodes is recommended. Depending on the stage of the tumor and the presence of metastases, adjuvant CTH is necessary for some patients [69, 70].

The unfavorable prognosis for survival increases from embryonal to pleomorphic to follicular forms. The prognosis in rhabdomyosarcoma depends on its stage. This neoplasm is curable in almost two-thirds of cases in the pediatric population. In contrast, pleomorphic forms in adults have a significantly worse prognosis. The spindle cell type in adults is characterized by an aggressive course [66–68].

Desmoplastic small round cell tumor

This is a tumor characterized by high malignancy and high grade already at the time of diagnosis. One of the few cases of a patient being cured of this tumor has been reported in the literature [71]. In the cited case of a 14-year-old boy, the tumor imitated an inguinal hernia. In the patient, the testis was not occupied by the tumor. The patient received 17 cycles of adjuvant CTH with vincristine, topotecan, cyclophosphamide, doxorubicin, etoposide, and ifosfamide after surgical treatment. In addition, the boy received 50.4 Gray of adjuvant irradiation to the tumor bed after the sixth cycle of chemotherapy. The boy was treated for 3 years and 1 month and remained on active oncological follow-up with no signs of local recurrence or distant metastases.
metastases. In that case, it was not specified whether the lesion had infiltrated the spermatic cord. However, it was a tumor of the perinuclear structures, which must be considered in the differential diagnosis of spermatic cord tumors [71].

Due to the rarity of this tumor, there are no established management guidelines. In our review of the literature, the preferred method of management for tumors in the inguinal region is to perform tumor resection, often with orchidectomy from inguinal access, with high resection of the spermatic cord. The standard of care for this tumor is neo- or adjuvant multidrug CTH (regimens are similar to those for Ewing sarcoma). On the other hand, RTH is proposed for patients who cannot undergo surgical resection [72].

Overall, 5-year survival in this tumor (for any location) is low at 15–20% [72]. Patients with a tumor location in the inguinal region have a better prognosis than patients with other tumor locations. This is because the tumor is more easily accessible, which increases its detectability. Patients can more easily palpate the lesion on self-examination and see a urologist more quickly.

In this group of patients, quoting from the authors of the cited article, 60% of patients were alive up to 120 months after treatment [71, 72].

Metastatic tumors

Metastasis of tumors from another location to the spermatic cord is extremely rare. Usually, the prognosis for patients with metastases to the spermatic cord is poor due to the mere presence of metastatic disease [73]. The primary tumor usually occurs in the gastrointestinal tract [73]. Metastases to the spermatic cord of tumors such as renal adenocarcinoma [1] and pancreatic adenocarcinoma [74, 75] have also been described in the literature. Treatment includes inguinal orchidectomy with high spermatic cord resection and postoperative chemoradiotherapy [75].

Conclusions

Tumors of the spermatic cord are generally benign. The most common benign tumor in this area is lipoma. However, the incidence of malignant tumors of the spermatic cord can be as high as 30% [76]. It is also worth noting that approximately 46% of soft tissue sarcomas are located in the thigh, buttock, and inguinal regions [77]. Therefore, it is recommended that careful differential diagnosis of tumors of the groin area should be performed, and that most lesions of this area should not be treated as inguinal hernia.

Author contributions

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