CASE REPORT

Paratesticular sarcomas: two cases with different evolutions

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SUMMARY

Paratesticular sarcomas are rare and account for less than 1% of all adult sarcomas. Intrasclerotical tumours can be testicular or paratesticular, paratesticular tumours being rarer (7–10%). Only 30% of paratesticular tumours are malignant and 90% of these are sarcomas. Histological subtypes include leiomyosarcoma, rhabdomyosarcoma, liposarcoma and undifferentiated high-grade pleomorphic sarcoma. Recurrence is frequent in this type of tumour and can occur years from initial diagnosis. These reports show two cases of paratesticular sarcoma with very distinct evolutions. The first case concerns a patient who presented with low-grade leiomyosarcoma with two local recurrences treated with surgery, and distance recurrence with cutaneous, subcutaneous, pulmonary and hepatic metastasis 30 years after surgery of the primary tumour. The second case reports of a patient who presented with high-grade myxoid liposarcoma with local and distance recurrence 3 years after surgery of the primary tumour, which progressed after chemotherapy; the patient died 7 months after diagnosis of recurrence.

BACKGROUND

Genitourinary sarcomas comprise less than 2.7% of all sarcomas. Paratesticular sarcomas account for less than 1% of all adult sarcomas. Scrotum tumours can have testicular or paratesticular origin.^{1 2} A paratesticular origin is rarer and represents only 7–10% of all intrascrotal tumours. These tumours can originate from the spermatic cord, epididymis or testicular tunica, however, frequently it is difficult to differentiate their exact origin. A total of 75% of paratesticular sarcomas originate from the spermatic cord.³ In total, 70% of paratesticular

Figure 2 H&E, Histological section of one of the subcutaneous nodules showing a pleomorphic fusocellular neoplasm that was positive to desmin and actin colorations compatible with metastasis of leiomyosarcoma (×40).

tumours are benign, being lipomas; adenomatoid tumours and leiomiomas are the most common histological types.⁴ Only 30% of paratesticular tumours are malignant and 90% of them are sarcomas.⁵ The histological subtypes include leiomyosarcoma, rhabdomyosarcoma, liposarcoma and undifferentiated high-grade pleomorphic sarcoma.^{6–9}

The most aggressive histological subtype is rhabdomyosarcoma, predominating in the first two decades of life; more differentiated tumours such as liposarcoma, leiomyosarcoma and fibrosarcoma are more frequently in adults.^{3 4}

Figure 1 One of the subcutaneous nodules, dispersed throughout the body, that was biopsied.

Figure 3 H&E, Histological section of one of the subcutaneous nodules showing a pleomorphic fusocellular neoplasm that was positive to desmin and actin colorations compatible with metastasis of leiomyosarcoma (×100).

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Figure 4 Chest, abdominal and pelvic CT scan showing multiple nodular pulmonary formations suggestive of secondary lesions (arrows).

When a scrotal mass presents, ultrasound, CT and MRI should be performed³ for the differential diagnosis and correct preoperative staging.

Treatment recommendations include radical orchiectomy, extended resection of the surrounding soft tissues and retroperitoneal lymphadenectomy in case of evidence of retroperitoneal lymph node metastasis or rhabdomyosarcoma, due to its high propensity to development of lymph node metastasis.³

Except for in patients with local recurrence and in high-grade rhabdomyosarcoma, adjuvant therapy with chemotherapy and radiotherapy showed little efficacy.³

Adequate treatment of local recurrence should include extended inguinal re-excision with radical orchiectomy, if not performed previously.⁶

Because of the high recurrence rate, prolonged follow-up is recommended and should include clinical observation, radiograms, CT and bone scintigraphy, if appearance of symptoms are suggestive of bone disease.³

The authors report two cases of paratesticular sarcoma with very different evolutions.

Figure 5 Chest, abdominal and pelvic CT scan showing multiple nodular pulmonary formations suggestive of hepatic lesions suggestive of secondary lesions, (arrows).

Figure 6 Chest, abdominal and pelvic CT scan showing multiple nodular pulmonary formations suggestive of a secondary mass lesion in the left anterior rectum muscle (arrows).

CASE PRESENTATION

Case 1

A 67-year-old man presented with a history of hypertension, dyslipidaemia and three surgeries to the left testicular mass 30, 25 and 17 years earlier, including left orchiectomy, of which the pathological diagnosis is unknown. He remained under surveillance and was asymptomatic. 15 years after his last surgery, the patient had noticed the appearance of subcutaneous nodules dispersed throughout his body. As the nodules increased in number over the next 2 years, he opted for a dermatology consultation. A biopsy of one of the lesions (figure 1) was performed and the pathology examination revealed a metastasis of leiomyosarcoma (figures 2 and 3).

The patient was referred to the oncology department. Clinically, he presented with a good general condition, with an Eastern Cooperative Oncology Group (ECOG) Performance Status (PS) of 0, with subcutaneous nodules dispersed throughout the body, without any other relevant alterations. Laboratory results did not reveal relevant alterations. A chest, abdominal

Figure 7 Revision of the lames of the testicular mass operated 17 years earlier at another hospital. Infiltration of the adjacent testicular tissue by the tumour (H&E).

Figure 8 Revision of the lamellae of the testicular mass operated 17 years earlier at another hospital. Detail showing high nuclear pleomorphism (H&E).

and pelvic CT scan showed multiple nodular pulmonary formations suggestive of secondary lesions (figure 4), hepatic nodules suggestive of secondary lesions with the biggest being 4 cm (figure 5) and a probable secondary mass in the left anterior rectum muscle (figure 6).

A biopsy of the abdominal wall lesion was performed and the pathological examination revealed a low-grade leiomyosarcoma, similar to the cutaneous lesion.

The pathology department of the hospital where the patient had undergone the 3 previous testicular tumour operations was consulted and his past records were accessed. A revision of the lamellae of the previous surgeries was performed and revealed a low-grade leiomyosarcoma of the spermatic cord (figures 7–9). Thus, we were in the presence of a patient with a leiomyosarcoma of the spermatic cord operated 30 years before, with two local recurrences treated by surgery, which now presented with cutaneous, subcutaneous, pulmonary and hepatic metastasis. Six cycles of chemotherapy with adriamycin and ifosfamide were performed on the patient; the disease stabilised and the patient remained under surveillance. A few months later, he presented a rapid disease progression with aggravation of his general condition and eventually died.

Figure 9 Revision of the lamellae of the testicular mass operated 17 years earlier at another hospital. Immunohistochemical staining showing immunoreactivity against actin.

Figure 10 Voluminous mass of the left hemiscrotum that presented with 3 years of evolution.

Case 2

A 67-year-old man presented with an enlarged left hemiscrotum with 3 years of evolution. For a recent increase of size, without any other symptoms, he addressed the hospital. The physical examination showed an enlarged left hemiscrotum (figure 10), without other relevant alterations. A scrotum echography was performed and revealed a voluminous homogeneous mass of the left hemiscrotum (figure 11). Chest, abdominal and pelvic CT scan excluded advanced disease and the patient underwent left orchiectomy with removal of a 17.6 cm paratesticular tumour weighing 1500 g, confined to the tunica vaginalis, without invasion of the testicle or epididymis (figure 12). The pathology examination revealed a high-grade myxoid liposarcoma (figures 13–15). The patient remained under surveillance and was asymptomatic, without evidence of recurrence. Thirty months after surgery, he noticed a left inguinal tumefaction. A abdominal and pelvic CT scan showed an abdominal mass with necrosis (figures 16 and 17) and pelvic lymphadenopathies (figure 18). A biopsy of the mass confirmed recurrence of myxoid liposarcoma and the patient started chemotherapy with

Figure 11 Scrotum echography confirming a voluminous homogeneous mass of the left hemiscrotum.

Figure 12 Surgical piece of the left orchiectomy presenting a 17.6 cm paratesticular tumour weighing 1500 g, confined to the tunica vaginalis without invasion of the testicle or epididymis.

adriamycin and ifosfamide. For clinical aggravation only three cycles of chemotherapy were administered, and the patient died of disease progression 7 months after detection of recurrence.

Figure 13 Pathological examination of the surgical piece of orchiectomy: lesion composed of myxoid matrix with delicate arborescent vessels. Differentiated areas of rounded cells and areas of necrosis can be observed. Lesion compatible with high-grade myxoid liposarcoma.

Figure 14 Pathological examination of the surgical piece of orchiectomy: signet ring small lipoblasts. Differentiated areas of rounded cells and areas of necrosis can be observed. Lesion compatible with high-grade myxoid liposarcoma.

Figure 15 Pathological examination of the surgical piece of orchiectomy: rare vacuolated lipoblasts on the periphery of the tumour. Differentiated areas of rounded cells and areas of necrosis can be observed. Lesion compatible with high-grade myxoid liposarcoma.

DISCUSSION

Paratesticular tumours are rare. However, recurrence is frequent in this type of tumour and can occur early (case 2) or many years after initial diagnosis (case 1). Thus, close follow-up is very important for early detection and correct treatment of recurrence.

Figure 16 Abdominal and pelvic CT scan showing an abdominal mass with necrosis, arrows.

Figure 17 Abdominal and pelvic CT scan showing an abdominal mass, arrows.

Figure 18 Abdominal and pelvic CT scan showing an abdominal mass with pelvic lymphadenopathies, arrows.

Although the role of adjuvant chemotherapy and radiotherapy is not well-defined (except in cases of high-grade rhabdomyosarcoma) and the benefit of survival is not proven, it should be considered and discussed on a case by case basis, at least in high-grade histology.³

Learning points

- ▶ Paratesticular sarcomas are rare tumours and account for less than 1% of all adult sarcomas.
- ▶ Intrascrotal tumours can be testicular or paratesticular, with testicular tumours being far more frequent (>90%) than paratesticular tumours (7–10%).
- ▶ Histological subtypes include leiomyosarcoma, rhabdomyosarcoma, liposarcoma and undifferentiated high-grade pleomorphic sarcoma.
- ▶ Treatment recommendations include radical orchiectomy, extended resection of the surrounding soft tissues and retroperitoneal lymphadenectomy if there is evidence of retroperitoneal lymph node metastasis or in cases of rhabdomyosarcoma, due to its high propensity to develop lymph node metastasis. The role of adjuvant chemotherapy and radiotherapy is not well-defined.
- ▶ Recurrence is frequent in this type of tumour and can occur many years after initial diagnosis. Close follow-up is very important for early detection and correct treatment of recurrence.

These reports show two cases of paratesticular sarcoma with very distinct evolutions.

The first case concerns a patient with low-grade leiomyosarcoma that presented two local recurrences treated with surgery, and distance recurrence with cutaneous, subcutaneous, pulmonary and hepatic metastasis 30 years after surgery of the primary tumour. The second case reports of a patient with high-grade myxoid liposarcoma, that presented with local and distance recurrence 3 years after surgery of the primary tumour, that progressed after chemotherapy with the patient dying 7 months after diagnosis of recurrence.

The earlier recurrence of case 2 is consistent with high-grade histology and is the habitual pattern of recurrence of sarcomas. On the contrary, the more indolent evolution of case 1, with 2 local recurrences treated with surgery and distant recurrence 30 years after initial diagnosis, is consistent with low-grade histology, and is an unusual pattern of recurrence of paratesticular sarcomas.

This article highlights the importance of close follow-up of patients with paratesticular sarcoma and the relevance of considering adjuvant therapy, at least for patients with high-grade histology.

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Competing interests None.

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