Leiomyosarcoma of the Inguinal Canal: A Rare Disease

Sir,

Leiomyosarcomas make up 5-10% of all soft tissue sarcomas in humans. The incidence within the inguinal canal is even lower, *i.e.*, <5%.[1] This has resulted in a dearth of guidelines regarding the proper approach and resolution. With less than 150 cases reported in the literature, the presentation of the leiomyosarcoma within the inguinal canal is a rare entity.[2] Peak incidence is usually reported within the 6th to 7th decades of life, with an increasing propensity for developing malignancy as the age of the patient progresses.[3] Herein, we present a case of a paratesticular leiomyosarcoma masquerading as an inguinoscrotal hernia.

A 46-year man presented to a urologist with a painless lump in the right inguinal region for 2 months. After baseline blood investigations and imaging, which revealed a localised mass in the right inguinal region, he underwent an excision of the lump. Histopathology revealed a high grade leiomyosarcoma of the right-sided mons pubis, 3.5x2.2x1.6 cm in size. On immunohistochemical staining, it was cytokeratin AE1/AE3 focal positive, ASMA diffuse positive, desmin diffuse positive, S-100 negative, and DOG-1 negative. The mitotic rate was 20/10 HPF. It was extended to an inked margin. The case was discussed in the tumour board meeting and decision was made to carry out wide local excision, including right orchiectomy. Therefore, he underwent wide local excision, including right orchiectomy. Histopathology did not reveal any evidence of malignancy in the entire surgical specimen. Postsurgery, he developed wound dehiscence, which was managed conservatively. Post-operative imaging on surveillance revealed no evidence of disease.

Preoperative differentiation between a benign or malignant growth within the inguinal canal is nearly impossible. Intrusive radiological investigations, such as CT or MRI, cannot reveal more than a large, complex, solid mass on imaging. This makes pre-intervention decision-making and realistic identification difficult. Definitive diagnosis is dependent upon the histological analysis of the specimen that can only be achieved after resection.

Due to the limited information available, standardised treatment options are limited. However, the consensus for radical orchiectomy with negative resection margins has been overwhelming. But this remains a double-edged sword, as attaining negative margins is nearly impossible due to the compact vascular anatomy of the inguinal canal, which increases the risk of inevitable loco-regional recurrence, *i.e.* (50% recurrence rate) without employing any add-on therapy.[4]

A number of adjuvant options have been utilised on a trial-and-error basis over the years, so as to effectively control this disease. In no particular order, radiotherapy, chemotherapy, and retroperitoneal lymph node dissection are a few regimens that have been adopted by clinicians.[5]

Chemotherapy, a viable option to prevent widespread haematogenous and lymphatic seeding, has its disadvantages. On the one hand, doxorubicin-based chemotherapy has proven itself to be an effective as well as the most widely utilised treatment modality for this particular disease process. However, few case reports and small-scale series in which it has been used, are not enough to establish its viability over other available management options. Further investigation is required before doxorubicin-centred chemotherapy can be considered as a concrete treatment option. SMAC conducted a meta-analysis in which 14 randomised control trials (RCTs) were evaluated, and it was established that although chemotherapy does, in fact, increase the margin for time to local and widespread failure, it is not by any great margin, with only a 4% absolute benefit being reported in a ten-year survival rate.[6]

On the other hand, there have been successful attempts at curbing local metastases by employing the use of radiotherapy, as reported by Fagundes *et al*.[7] They deduced that there was a reduction of local recurrence from 50% to 10-20%. However, there is not enough data to support its standardisation. Other authors hold the opinion that it has no role to play in controlling the recurrence of the disease, as metastases are mostly haematogenous and therefore immune to the effects of irradiation.[3]

Whatever modality is employed to target and eradicate the disease, it is essential to provide the patient with long-term surveillance.

COMPETING INTEREST:
The authors declared no competing interest.

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