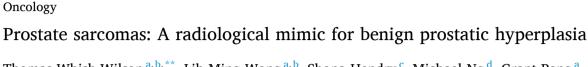
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ARTICLE INFO	A B S T R A C T
<i>Keywords:</i> Sarcoma Prostate cancer MRI Leiomyosarcoma	Leimyosarcomas arising from the stroma of the prostate are very rare, accounting for 0.1% of malignancies. We describe a case that closely mimicked benign prostatic hypertrophy on magnetic resonance imaging. Due to the low incidence of disease there is no high level evidence for management. We advocate neoadjuvant radiotherapy followed by radical prostatectomy with pelvic lymph node dissection. Diagnosis and expedient management is critical.

Introduction

Leiomyosarcoma of the prostate is a rare malignancy accounting for 0.1% of prostate malignancies.¹ It was first described in 1853 and English literature searches show case reports from 1950 onwards, since then there have been less than 200 cases reported globally.² Although rare, clinicians must be aware of this aggressive malignancy with a median survival of 17 months and 5-year survival rates of 26%.^{2,3} They effect male patients of all ages from 2 to 80 years.^{2–5}

Case presentation

A 58 year old man with no significant past medical history presented to his local doctor with exacerbation of pre-existing lower urinary tract symptoms, most notably increased frequency and nocturia. Previous investigations, 4 years earlier, were unremarkable. He was an active man who took no regular medication, was a non-smoker, and had no surgical history.

Prostate specific antigen (PSA) was 2.2ng/ml, creatinine 85 µmol eGFR 87ml/min/1.73m². Ultrasound studies showed prostatomegaly (62 cc), and new elevated post micturition volume of 108ml with a thickened trabeculated bladder. Significantly, the prostate demonstrated a hypoechoic solid lesion arising from the right-side of the gland measuring $24 \times 26 \times 23$ mm, which was not present 4 years earlier

(Fig. 1).

The patient was referred to an urologist who performed a digital rectal exam which was benign. Multi-parametric magnetic resonance imaging of the prostate was obtained, revealing a right-sided paraprostatic nodule $(2.9 \times 2.1 \times 2.5 \text{ cm})$. The lesion was in contact with the bladder and obturator internus muscle with no invasion. It was heterogeneous and on T2 imaging resembled identical morphology to the hypertrophied transition zone, with presumed stromal and glandular elements within (Fig. 2a). However, on the high B value DWI, there was elevated signal within the stromal elements and restricted signal on the ADC to around 800 (Fig. 2b). The enhancement is heterogeneous on the DCE similar to the transition zone. There was no targetable lesion within the prostate. There was no extracapsular extension or abnormal pelvic nodes.

A CT-guided biopsy was performed and three 18-gauge core biopsy were obtained. All cores comprised of entirely lesional material with no prostatic tissue observed. Cores were composed of highly atypical spindle cells arranged in intersecting fascicles. The cells had elongated, hyperchromatic, pleomorphic nuclei and abundant densely eosinophilic cytoplasm. There were numerous apoptotic nuclei and mitotic figures seen, numbering up to 2 per 10HPF. There was no tumour necrosis.

Cells were strongly and diffusely positive for SMA and desmin. CD34 showed patchy positivity. S100 and SOX-10 were negative. The Ki-67 proliferative index was variable, up to 20% in the highest areas.

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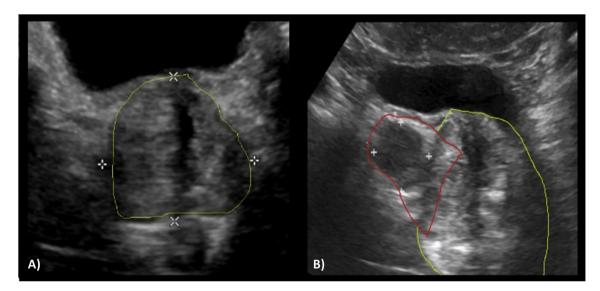


Fig. 1. Ultrasound transverse view of prostate (green line) A) 07/08/2015, B) 25/06/2019 with new lesion arising from right lobe (red line). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

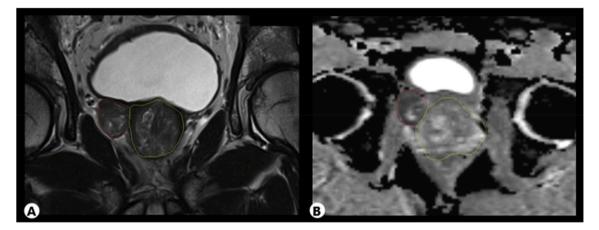


Fig. 2. MRI axial views with prostate in green and lesion in red A) T2, B) ADC. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

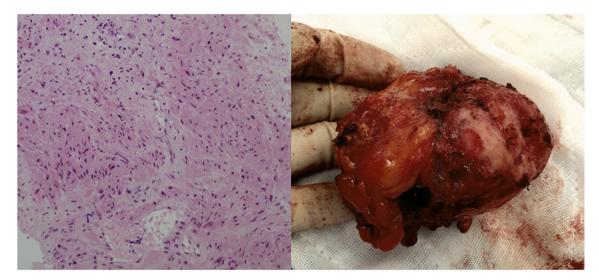


Fig. 3. Haematoxylin and Eosin stain $10 \times$ (left); Anatomical pathology specimen viewed from right lateral with lesion on left of picture and apex of prostate on right of picture (right).

A computed tomography (CT) scan of the patient's chest and FDG-PET scan was performed for staging, which revealed no avid visceral or nodal metastases and moderate grade peripheral metabolic activity in the leiomyosarcoma. The patient was referred to a dedicated sarcoma multidisciplinary meeting. The decision for neoadjuvant radiation therapy (50.4Gy, 28 fractions) followed by a robotic assisted radical prostatectomy (RARP) was decided. Following radiotherapy subsequent imaging showed interval necrosis of the tumour, which remained PETavid. RARP and right pelvic lymph node dissection was performed five weeks post completion of radiotherapy, which was unremarkable, adhering to minimal tissue handling with wide margins (Fig. 3).

Final histology showed a 58mm tumour, which was upgraded from the original biopsy from grade 2 to grade 3 leiomyosarcoma. Margins were clear with no infiltration of seminal vesicles, fat planes present, and a medial margin formed by the prostate with no infiltration. Nodes were reactive with no metastases detected.

Discussion

Although rare, accounting for less than 0.1% of prostate malignancies clinicians need to be mindful of this aggressive disease.⁵ Prostate leiomyosarcomas are a subgroup of sarcomas affecting the prostate, which include rhabdomyosarcomas, carcinosarcomas, and unspecified.

Leiomyosarcomas are difficult to identify on MRI as they resemble benign prostatic hypertrophy (BPH) on T2 imaging and heterogeneously enhance on DCE much like the transition zone. This may make intraglandular prostatic lesions difficult to identify. However, on the high B value DWI, there is elevated signal within the stromal elements and restricted signal on the ADC to around 800 reflecting the hypercellularity of these tumours, making them somewhat suspicious. This coupled with a low PSA may delay diagnosis contributing to poor outcomes. MRI for evaluation of patients with suspected prostate cancer has become standard of care and is supported by a large body of evidence. If the lesion had been confined to the transition zone, based upon PIRADS version 2 it would have been classified as a PIRADS 2 lesion and avoided biopsy. In PIRADS version 2.1 the role of DWI in the transition zone has been expanded and would have upgraded this lesion to a PIRADS 3. However given the low PSA the patient would still be unlikely to have undergone biopsy. We were fortunate with this patient given the

exophytic nature of his lesion.

Most cases have been treated with external beam radiotherapy as more common sarcomas are known to be radiosensitive, however, there is not enough literature to support this and treatment is fiduciary and empirical.⁵ Despite the prompt detection and subsequent management this patient's sarcoma was upgraded on final histology, which highlights the aggressiveness and timely manner this tumour must be managed. Published survival outcomes are poor but with a 17 month median but it is hoped our patient will fare better. We have proposed annual follow up with FDG-PET alternating with MRI. At the time of writing he is six months from original biopsy, and recovered well from his treatment.

Conclusion

Leiomyosarcomas of the prostate are rare. They often present with non-specific urinary tract symptoms, most commonly increased urinary frequency. Unless exophytic, diagnosis remains challenging on imaging and delay may be increased due to normal PSA levels and standard investigation algorithms. Due to the rare nature of this disease there is no consensus on best management approach. We recommend consultation of experienced sarcoma teams for guidance. This case will add to the literature so that an eventual treatment algorithm can emerge.

Declaration of competing interest

The authors have no affiliations that may prejudice this report or any relevant conflicts of interest.

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