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CASE REPORT

Primary prostate sarcoma: how to manage following diagnosis at transurethral resection

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Abstract

Primary prostate sarcomas are rare, reportedly comprising just 0.7% of all prostate malignancies. Here, we present the case of a 66-year-old man who was diagnosed with prostate stromal sarcoma after undergoing a routine transurethral resection of prostate for bladder outflow obstruction. Primary prostate sarcoma can be aggressive even when low-grade, with a high risk of local recurrence and, high malignant potential when high-grade. They require aggressive multimodality treatment with surgery, chemotherapy and radiotherapy for durable survival outcomes. They also require close surveillance with long-term follow-up.

INTRODUCTION

Primary prostate sarcomas are rare, with one study estimating it to comprise of just 0.7% of prostatic malignancies [1]. It typically arises from specialized hormone-dependant mesenchymal cells of the prostatic stroma [2]. A diverse histological pattern has been described in the literature, often closely related to stromal tumours of uncertain malignant potential (STUMP). Here, we describe the case of a primary prostate stromal sarcoma in a patient following transurethral resection of prostate (TURP).

CASE REPORT

A 66-year-old man initially presented with low-pressure, acute urinary retention. He was catheterized and commenced on an alpha-blocker (tamsulosin). Initial assessment of the patient's prostate by digital rectal examination noted it to be enlarged and benign-feeling. After 2 weeks, he had an unsuccessful trial without catheter and was consented for a TURP. During TURP, he was noted to have abnormal-appearing and chalk-white tissue within the left prostatic lobe on resection; the hardness of the resected tissue was such that it damaged the resecting bipolar loop twice.

Malignancy was suspected but the histology report returned surprisingly as demonstrating high-grade prostatic stromal sarcoma, in which 80% of the 36 grams of TURP chippings contained malignant, spindle cell proliferation without an organized fascicular pattern but with the presence of nuclear atypia, mitotic activity and tumour necrosis (Figs 1–3).

Multiparametric magnetic resonance imaging (MRI) of the prostate and pelvis demonstrated a heterogeneous enhancement of the remaining prostate tissue. The prostatic capsule was intact; however, there were enlarged left iliac lymph nodes (Figs 4 and 5). Staging computed tomography (CT) of thorax, abdomen

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Figure 1: Macroscopic sections of the apical, mid-gland and basal areas of the prostate specimen demonstrating central TURP cavitation and abnormal residual tissue



Figure 4: T1-weighted axial MRI image with contrast demonstrating an enlarged prostate with inhomogenous signal



Figure 2: Haematoxylin and Eosin staining with $\times 10$ magnification demonstrating ovoid, short spindle cells without an organized growth pattern



Figure 3: Haematoxylin and Eosin staining with \times 40 magnification demonstrating hypercellularity, nuclear hyperchromasia and mitotic activity



Figure 5: T2-weighted axial MRI image with contrast demonstrating an enlarged prostate with inhomogenous signal

and pelvis did not show evidence of progression elsewhere in the body. The patient therefore underwent an open radical cysto prostatectomy with retroperitoneal lymph node dissection and urinary diversion.

DISCUSSION

Primary prostate sarcomas are rare. The initial presentation can be with features of bladder outflow obstruction (BOO) such as lower urinary tract symptoms or acute urinary retention, haematuria or an abnormal finding on digital rectal examination [3–6]. BOO is typically reported as being the most common presentation [3–6]. Prostate-specific antigen may be elevated but is typically normal [4]. It has also been described following pelvic radiotherapy [4].

Histologically, the stroma within the prostate gland contains specialized hormonally responsive cells, which participate in complex stromal-epithelial interactions. It has been postulated that the exaggeration of these interactions may lead to the development of primary sarcoma [6]. Primary prostate sarcoma is histologically characterized by stromal hypercellularity, nuclear atypia, mitotic activity and necrosis [7]. The histological pattern is diverse with stroma cells varying from round and plump to spindled-shaped [6]. It can be classified as low- and high-grade. High-grade prostatic sarcoma histologically shows proliferation of epithelioid and spindle cells that show an abnormal pleomorphic and hypercellular growth pattern with marked nuclear atypia, mitosis and necrosis. Low-grade disease, in comparison, has no nuclear atypia and lower mitotic rates [2, 4]. Sarcomatous subtypes are mainly leiomyosarcoma and rhabdomyosarcoma [5, 8]. Primary prostate sarcoma has also been closely linked to STUMP, which is a stromal proliferation of the prostate with a variety of patterns described [7]. It is closely associated with primary prostate sarcoma because it has been described both concurrently and metachronously [4, 6, 7].

Radiologically, prostate sarcoma appears as a heterogeneous mass with rapid hypervascular and heterogeneous enhancement with contrast on CT and MRI. Cystic areas may also be present [5].

The management of primary prostatic sarcoma consists of a combination of radical surgery, radiotherapy and chemotherapy, in which different combinations have been used [3, 9]. Surgery alone is not deemed adequate for long-term survival. Long-term survival, as well as freedom from local recurrence, is associated with clear surgical margins and the absence of metastases at diagnosis [8]. Age of >50 years, metastases and lack of surgery with curative intent are related to poor prognosis [1]. Aggressive treatment is required as local recurrence is common, even in low-grade disease [4, 10]. Recurrence has been described as occurring in up to 100% of cases in with high-grade tumours [10]. High-grade tumours also have a high metastatic potential [2]. They most commonly metastasize to the lungs and bones [10]. One series reported that 62% of patients had distant metastases at diagnosis [8]. Five-year overall survival ranges from 11.3% to 38% [1, 8].

In conclusion, primary prostatic sarcoma is a potentially aggressive disease even when low-grade. The likelihood of local recurrence and metastases warrants that patients with this diagnosis undergo prompt radical surgery with adjuvant or multimodal treatments subsequent to which close surveillance with long-term follow-up is required.

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CONFLICT OF INTEREST STATEMENT

None declared.

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