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Introduction

Bladder paraganglioma accounts for only 0.06% of bladder tumours¹. Paragangliomas are chromaffin tumours or extra-adrenal phaeochromocytomas which can be found anywhere along the paravertebral and para-aortic axes.² Prostatic paraganglioma is exceptionally rare with only 11 cases reported to date.³ Amongst reported cases, those which have been managed surgically were treated with radical prostatectomy.⁴ To our knowledge, this is the first reported case of partial or anterior prostatectomy for bladder paraganglioma involving the prostate.

We discuss a case of successful partial prostatectomy for the excision of bladder paraganglioma with prostatic invasion using an open retropubic approach.

Case report

A 19-year old male with known familial paraganglioma (heterozygous germline mutation in SDHB gene) presented to hospital following a low-speed motorbike accident. Significant medical history included a diagnosis of multiple paraganglioma 18-months prior, requiring right

nephroureterectomy, partial cystectomy, and resection of aortocaval and para-iliac masses. Continued surveillance deemed the patient clinically well until this presentation to hospital.

A full trauma work-up led to an incidental finding of a new low-lying intravesical mass suspicious for recurrence of paraganglioma. The patient was asymptomatic for signs of adrenal excess. He denied previously encountered symptoms of haematuria or micturition syncope. Biochemical testing showed elevated serum normetanephrines and methoxytyramine confirming functional paraganglioma.

Pre-op clinical staging comprised CT, MRI, bone scan and (⁶⁸Ga)DOTA-TATE (GaTate) PET nuclear scan. No metastatic disease was found. MRI showed a 4.8cm lesion at the anterior aspect of the prostate, abutting the prostatic urethra, with a large feeding vessel and left-sided enhancing nodes (Figure 1). MIBG scan did not demonstrate any uptake.

Treatment options included intra-operative brachytherapy, but was considered by the multidisciplinary team to be too inhomogenous; radiotherapy via consolidative external beam radiotherapy, however delivery of a safe dose post-operatively would be limited; cytoreductive systemic therapy; or, cysto-prostatectomy with node dissection and conduit/neobladder construction if required, with or without neoadjuvant radionuclide treatment. Consensus recommendations were for prostatectomy and/or cystectomy. The aims of surgery were improvement of functional outcomes as well as disease control.

Open excision of the paraganglioma tumour via anterior partial prostatectomy with left pelvic lymph node dissection was undertaken. A midline lower abdominal incision was made. Extensive adhesiolysis was performed with careful dissection into the retropubic space. The endopelvic fascia was divided bilaterally and the prostate dissected out to its apex. The bladder neck was opened anteriorly. A well circumscribed tumour of the anterior bladder neck and prostate was found and resected completely. (Figure 2) There was no involvement of the posterior portion of the prostatic urethra allowing for partial preservation. The dorsal venous complex was over-sewn at the prostatic

apex. An anastomosis was created between the anterior bladder neck flap and the posterolateral prostate with good neurovascular bundle preservation.

Histopathology sections showed a 40mm tumour centred in the muscularis propria of the bladder wall, with focal invasion of perivesicular fat, skeletal muscle and prostatic parenchyma. Tumour margins were transected at 3 and 9 o'clock. Nests of cuboidal cells with granular eosinophilic cytoplasm and pleomorphic nuclei with chromatin clearing were seen. Tumour cells strongly expressed synaptophysin, chromogranin and CD56. S100 stained sustentacular cells. (Figure 3) Left inguinal and obturator nodes did not demonstrate tumour.

Intra-operative and post-operative stages were uneventful with normal urogenital function and without signs of functioning paraganglioma. The patient received adjuvant Lutate therapy for 6-months post-operatively. At 2-year follow-up, no tumour recurrences were seen on MRI, and known nodal lesions remained stable on nuclear PET; plasma free metanephrines remained within the normal range. This confirmed successful functional outcomes without disease progression.

The final diagnosis was bladder paraganglioma with focal prostatic invasion.

Discussion

The first reports of partial prostatectomy for infiltrating carcinoma of the bladder neck was a case series published in 1929.⁵ Other reports in the literature relate to rectal cancer invading the prostate. More recently, robotic-assisted partial prostatectomy has been described in a recent surgical series of 17 patients for cases of anterior prostate cancer.^{6,7} Although controversial in the context of prostate cancer,⁷ the treatment of choice for localized and solitary paraganglioma involves complete surgical resection of the tumour under adrenergic blockage.^{2,4,8}

To our knowledge, this is the first report of partial prostatectomy for focally invasive paraganglioma. This case demonstrates a rare example of bladder paraganglioma with prostatic invasion, where partial prostatectomy was safe and feasible in the right patient setting. Although it carried some risk of margin positivity, overall disease progression was unaffected whilst maximally preserving

functional outcomes. Multidisciplinary consultation between surgical, neuroendocrinological, oncological, and medical imaging specialists were pertinent to successful outcomes achieved. Continued surveillance is paramount in young persons with familial paraganglioma.

Disclosure statement

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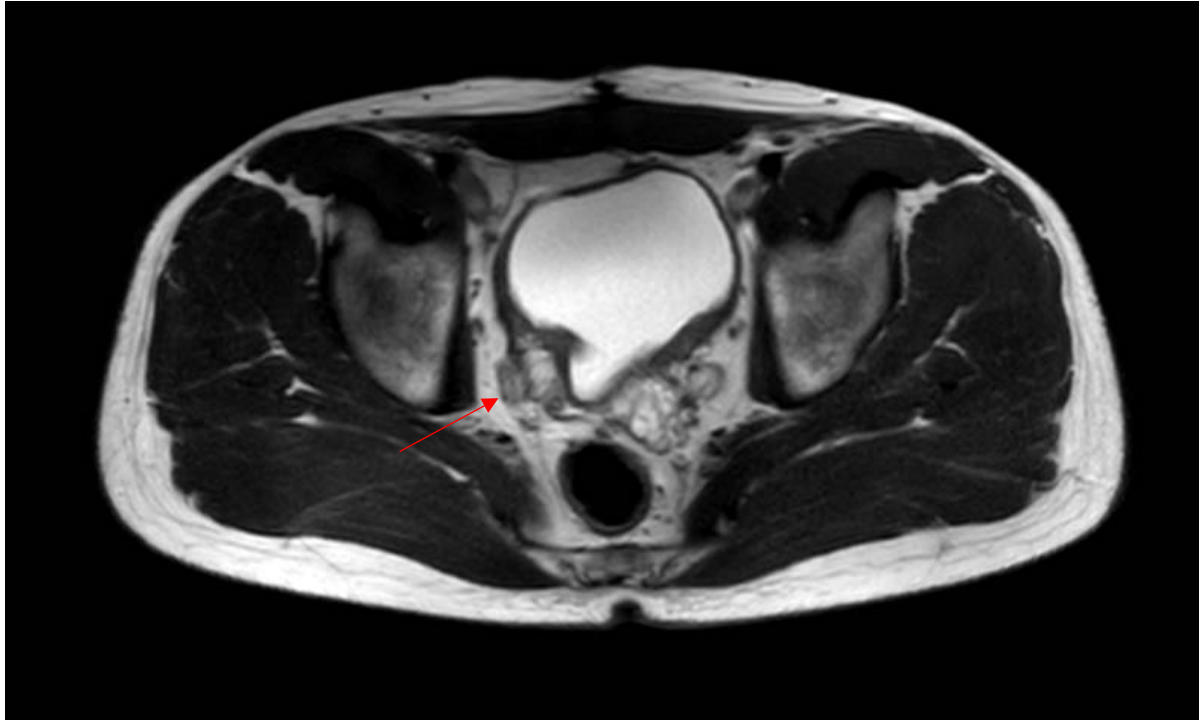
Figure Legend

Figure 1: MRI (T2 weighted) Axial view

Figure 2: Pathology

Figure 3: Histopathology

Figure 1: MRI (T2 weighted) Axial view with gadolinium contrast



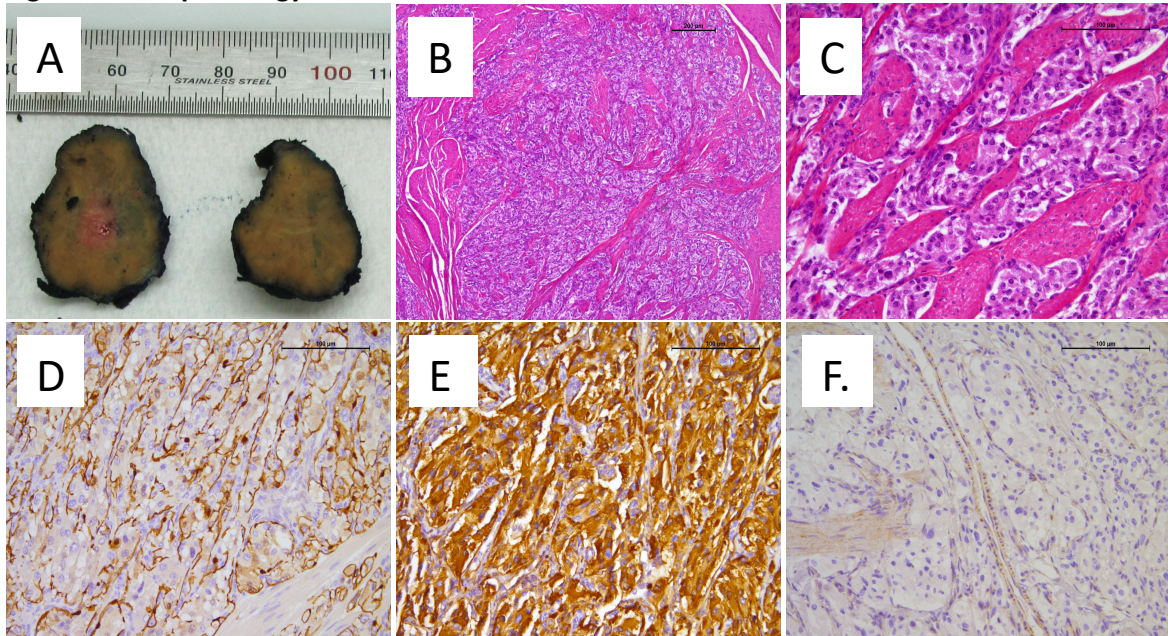
MRI confirming a solid lesion in the anterior aspect of the prostate abutting the prostatic urethra.

Figure 2: Pathology



The gross specimen weighed 26g and had dimensions of 44x31x25mm. The cut surfaces were firm tan-brown and homogeneous.

Figure 3: Histopathology



A. Cut surfaces of gross specimen. B&C. Haematoxylin and eosin stained sections of tumour. D-F. Immunostains for S100, chromograinin A and SDHB.