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Solitary extramedullary plasmacytoma of prostate on the background of treated prostate adenocarcinoma

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ABSTRACT

Solitary Plasmacytoma is a rare entity which accounts for < 5% of all plasma cell dyscrasias. Most commonly involved sites for solitary extramedullary plasmacytoma are head and neck (90 %) especially upper respiratory tract, followed by Gastrointestinal tract. Rarely it can involve testes, bladder and orbit. PET/CT provides an invaluable imaging support for initial evaluation of additional plasmacytomas and response to treatment. There are no randomized trials regarding the best treatment approach. Generally, radiotherapy remains treatment of choice of solitary plasmacytoma. Our case, with its rare site of occurrence along with patient's history of radiotherapy treated prostate malignancy and significant history of radiation cystitis provides a unique diagnostic and therapeutic challenge. Treatment can be personalised in cases such as this, where patients can be treated with chemotherapy in contrary to radiotherapy as further exposure to the radiations could exacerbate the bleeding risks.

Keywords: extramedullary plasmacytoma of prostate , prostate plasmacytoma

Contributions: RF wrote the paper. AH reviewed and corrected the manuscript. RF and AH treated and managed the patient.

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Introduction:

An 83 year old man presented in emergency department with recurrent episodes of painless frank hematuria. He had a history of radiotherapy treated prostate adenocarcinoma (2003) and multiple myeloma diagnosed 4 years ago, which was in Complete remission (CR). His other co-morbidities included chronic obstructive pulmonary disease, ischemic heart disease, hypertension, type II Diabetes mellitus and transient ischemia attack.

He was diagnosed with prostate adenocarcinoma 17 years prior to this presentation and was successfully treated with radiotherapy (70 Gy in 35 fractions) as he was deemed unfit for radical prostatectomy that time. Four years prior to this presentation with hematuria, he was referred to haematology service with multiple ribs osteolytic lesions and IgA monoclonal band of 21g/l. Bone marrow biopsy confirmed the diagnosis of multiple myeloma, ISS stage I, with plasma cells accounting for 70% of cellularity of trephine. Cytogenetics revealed high risk profile with gain(1q). He was initiated on Lenalidomide and dexamethasone. Treatment with lenalidomide and dexamethasone was continued for 18 months and stopped due to lack of acceptability of this treatment to the patient. At the time of stopping his treatment his myeloma was in CR. Three years later, he presented in the emergency department with recurrent episodes of haematuria. Initial investigation with CT-urogram and cystoscopy showed moderately enlarged, vascular and malignant looking prostate. Initial preliminary diagnosis of Prostate adenocarcinoma was made with significant background changes of radiation cystitis. His haematuria was managed by continuous bladder irrigation and he was discharged home with catheter in situ once bleeding settled and was placed on hormonal therapy. This was followed by further three hospital admissions with similar presentations which were temporarily managed by intravesical sodium hyaluronate therapy, diathermy and bladder

washout. Subsequently, rigid cystoscopy was performed, which revealed bleeding hyper-vascular prostate. Multiple fragments of haemorrhagic tissues (prostate chips) were sent for histological analysis and later embolisation was performed which failed to stop bleeding.

Histology revealed prostate tissue diffusely replaced by a population of pleomorphic cells with the staining characteristics of plasma cells (CD138+, Kappa+), with Kappa restriction. Immunohistochemistry was negative for markers of small cell differentiation, prostate, urothelial and keratin markers. Serum protein electrophoresis showed no monoclonal band and normal serum light chains with no evidence of end organ damage. Patient refused bone marrow biopsy. Positron Emission Tomography – Computed Tomography (PET-CT) revealed intensely FDG(Fluorodeoxyglucose) avid mass in the prostate gland extending into the bladder and left perineum (SUV max 16). No other foci of increased FDG uptake was noted. (See Figure a)

Diagnosis of Solitary extramedullary plasmacytoma was established and patient was started on CyBORd. (Cyclophosphamide 350mg/m² weekly, Bortezomib 1.3mg/m² day1,8,15,22 and Dexamethasone 20mg weekly). He tolerated treatment very well. His haematuria resolved completely during the first cycle and was discharged from the hospital. He continued to receive CyBORd in Haematology Day ward. A repeat PET-CT was performed to assess treatment response after 2 cycles, which showed no residual FDG avidity in prostate consistent with excellent response to treatment (see Figure b). Patient is due to start 4th cycle of the treatment and is followed up monthly in day ward.

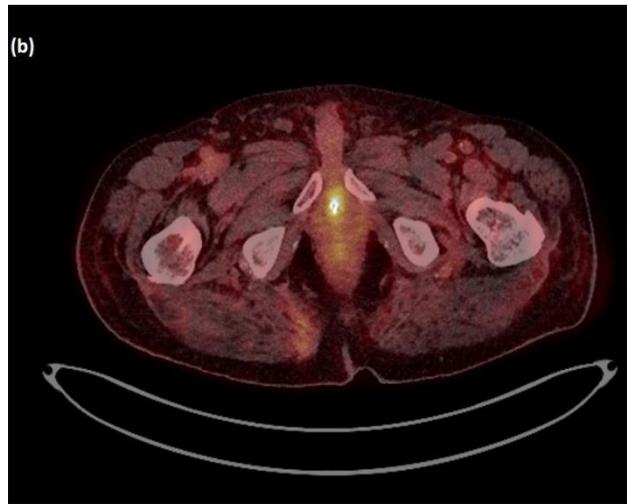
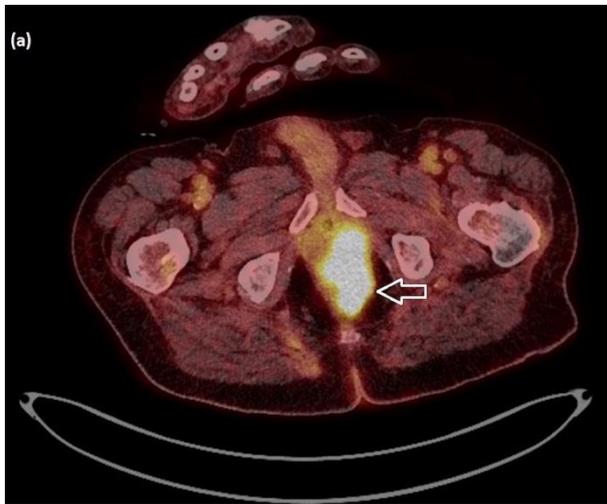
Discussion:

Solitary plasmacytoma (SP) is characterised by localised proliferation of neoplastic monoclonal plasma cells. It is a rare entity which accounts for < 5% of all plasma cell dyscrasias.¹ Depending on the site of occurrence, it can be broadly classified as Solitary Plasmacytoma of

Bone (SBP) and Solitary Extramedullary Plasmacytoma (SEP). SEPs are less common than SPBs. It has a male: female ratio of 3:1 and median age of diagnosis is 55 years.¹

Most commonly involved sites for SEP occurrence (90 %) are head and neck especially

upper respiratory tract, followed by Gastrointestinal tract. Rarely it can involve testes, bladder, orbit.^{2,3,4,5} Previously one case of prostate involvement is reported along with systemic disease (active myeloma).⁶



(a) Pre-treatment PET-CT ; Intensely FDG-Avid mass (shown by arrow) in the prostate gland extending into the bladder and into the left perineum (SUV max 16) (b) Post cycle 2 PET-CT scan; No FDG avid mass in the prostate

As per Revised IMWG criteria, diagnosis of solitary extramedullary plasmacytoma requires biopsy proven solitary lesion of soft tissue with evidence of clonal plasma cells, a normal bone marrow with no evidence of plasma cells, no end organ damage (CRAB features) and no evidence of bony disease on imaging. PET-CT continues to be an invaluable imaging modality for initial evaluation of SP and treatment response. Previously MRI was recommended to assess solitary bone plasmacytoma and to exclude other additional lesions. Using PET-CT, Schirrmester et al (2003), detected additional lesions in 33% of patient with SP when compared to standard imaging.¹⁰ Similarly, Nanni et al (2008), detected additional lesion in 43% patient which were missed with standard imaging including MRI and plain X-Rays.¹¹

There are no randomized studies regarding the best treatment approach. Radiotherapy is the treatment of choice for SP. however its efficacy has been tested only in small

retrospectivestudies.^{7,8} One study by Goyal et al, largest retrospective study covering solitary plasmacytomas analyses different treatment options including surgical approach. Combined RT and surgery improved survival compared to either modality alone, but whether the survival differences are due to treatment given or the site involved, is not known.⁹

Our case above is an extremely rare occurrence as no solitary extramedullary plasmacytoma of the prostate has been reported in the past. Its site of occurrence along with the patient's history of radiotherapy treated prostate adenocarcinoma and significant background changes of late radiation cystitis provides a very unique diagnostic and therapeutic challenge. Radiological and cystoscopic findings provided a compelling evidence to the physicians to treat the patient for a primary prostate malignancy with hormonal therapy. Hence, it signifies the importance of biopsying the prostate for a definite diagnosis. Moreover, a personalised

approach should be used in certain cases such as this, where the patients can be treated with chemotherapy in contrary to first line management with radiotherapy as further exposure to the radiations could possibly exacerbate the bleeding risks.

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