Neurofibromatosis mimicking perineural invasion in prostate adenocarcinoma

Clinical History:

A known case of neurofibromatosis 1 (NF1), the patient was diagnosed with prostate adenocarcinoma (PCa) on biopsy (Gleason Score 4+3=7 in 7/12 cores, highest volume 50%, without perineural invasion).

Multiple neurofibromas were noted on eyebrow, neck, head, and gluteal fold.

Family history: PCa in father; NF1 in 2 offsprings.

Imaging Findings:

The MRI, in addition to findings compatible with the biopsy-proven PCa, revealed an ill-defined area of enhancement along the left pudendal nerve extending through the sciatic notch (Figures 1a-c). This enhanced lesion measured approximately 3.8 x 8.0 cm in antero-posterior and transverse dimensions respectively and was concerning for perineural spread, therefore the findings were interpreted as EPE involving the right apex.

A biopsy of the enhanced mass in the sciatic notch was obtained later and revealed a neurofibroma.

Bone scan was negative for metastatic disease.

Discussion:

Perineural invasion (PNI) in prostate adenocarcinoma (PCa) highlights increased risk for extraprostatic extension (EPE), seminal vesicle invasion and shorter disease-free interval; thus advocating more radical intervention (non-nerve sparing surgery and hormone therapy) [1, 2].

MRI is a useful tool in identification of PCa where T2 signal hypointensity with early enhancement and diffusion restriction is noted [3]. MRI can detect high grade tumour more readily than low risk PCa [4]. The majority of PCa are peripherally located, and stage of tumour is based on confinement to the prostate. Non-organ-confined disease consists of seminal vesicle invasion, invasion or asymmetry of the neurovascular bundles, obliteration of the recto-prostatic angle and bulging of the prostate capsule [5]. In distinction, perineural spread of PCa on multiparameter (MP)-MRI appears as infiltrative pattern, which can be demonstrated as fusiform, nodular or thickened periprostatic nerves [6]. Diffusion-weighted MRI is suggested to improve the prediction of EPE [7].

Gordetski et al [8] reported improved PNI detection using MP-MRI in 64 cases. Using MP-MRI and MRI/ultrasound fusion guided biopsy, the authors detected PNI in 9 additional cases of 19 not diagnosed by standard biopsy; however, they missed 2 PNI cases diagnosed by standard ultrasound-guided biopsy. In their cohort, PCa was missed altogether by MP-MRI in one case with scattered disease.

Neurofibromatosis type 1 (NF1) is a rare disease where mutation in Neurofibromin (chromosome 17) elicits tumours
of the nervous system. Pelvic involvement is uncommon in NF1, mainly affecting the bladder in children and youths [9]. The best diagnostic imaging modality for pelvic neurofibroma is MRI. Localized and plexiform neurofibromas typically show 'target' appearance with peripheral T2 hyperintensity and T2 central hypointensity [10]. Following contrast administration, there may be a reverse target-shaped enhancement.

In our case, the history of NF1 was not available to the radiologist at the time of initial MRI interpretation; and EPE was reported based on bulging of the prostate capsule along the posterior aspect of the prostate and possible perineural spread based on the enhancement and thickening along the pudendal nerve. This resulted in overtreatment (non-nerve-sparing rather than nerve-sparing RP) with higher potential for complication. But RP revealed organ-confined PCa with neurofibromas in the posterior neurovascular bundles causing indentation on the prostate.

MRI is helpful in the diagnosis of PCa, but its administration has limitations. Pelvic neurofibromas can mimic PNI and EPE. Obtaining adequate clinical information is very important.

**Differential Diagnosis List:** Prostate adenocarcinoma, organ-confined (pT2b N0) with pelvic neurofibromas., Extraprostatic extension of prostate adenocarcinoma, Neurofibromatosis of pelvic organs

**Final Diagnosis:** Prostate adenocarcinoma, organ-confined (pT2b N0) with pelvic neurofibromas.

**References:**


Description: (Axial T2 non-fat-saturated image) Decreased T2 signal at the right aspect correlating to PCa (black arrow). The circumscribed homogeneously T2 hyperintense mass (white arrow) correlates with neurofibroma. Origin: Amin A, Brown University, Providence, RI
Description: (ADC map) Area of diffusion restriction at the right apex correlates to PCa (black arrow). The circumscribed mass without diffusion restriction correlates with neurofibroma (white arrow). Origin: Amin A, Brown University, Providence, RI
**Description:** (post-gadolinium fat-saturated T1 axial) Enhancement along the left sciatic notch, initially interpreted as perineural spread (white arrow). Biopsy revealed a plexiform neurofibroma. **Origin:** Amin A, Brown University, Providence, RI

**Description:** Neurofibroma causing mass-effect on the prostate in the right posterior apical segment. (H&E, 10X) **Origin:** Amin A, Brown University, Providence, RI