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Primary prostate solitary neurofibroma without neurofibromatosis-I: a case report and narrative review of the literature

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► ***Case report***

ABSTRACT

Neurofibroma of the prostate is an extremely rare benign tumor, particularly when it is not associated with neurofibromatosis-I (NF-1). This study aimed to report the case of a 44-year-old man who presented with dysuria for about 4 years. Magnetic resonance imaging (MRI) showed a mass in the prostate with "target sign" on T2-weighted imaging. The mass was surgically excised, and histopathological findings revealed a neurofibroma. No clinical or laboratorial manifestations of NF-1 were identified. Also, no local recurrence occurred after the 3-month follow-up. This was the first case of prostate solitary neurofibroma without NF-1, and the "target sign" on MRI could supply some significant implications in the diagnosis.

Keywords: *Neurofibroma, Prostate tumor, Solitary, Magnetic resonance imaging.*

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INTRODUCTION

Neurofibroma is a benign tumor of the nerve sheath originating from Schwann cells. It may present individually or as part of neurofibromatosis-I (NF-1) [1]. Neurofibromas are commonly seen in the neck, thorax, cranium, retroperitoneum, and upper and lower extremities. However, neurofibromas arising from deep organs are rarely reported. Pelvic neurofibroma is a rare entity, with around 60 cases reported so far [2]. Prostatic neurofibroma is even rare, and only four cases of neurofibroma of the prostate with NF-1 have been reported so far [2-4]. Table 1 summarizes the pertinent characteristics of these patients. This study aimed to report the first case of prostate solitary neurofibroma without NF-1 and the role of magnetic resonance imaging (MRI) in its diagnosis.

Case report

A 44-year-old man presented with dysuria and fullness in the lower abdomen for about 4 years. He could not self-void for about 1 month. The ultrasound examination revealed benign prostatic hyperplasia (7 × 5 × 3 cm^3). The patient did not receive any treatment 3 years ago. The mass felt big and hard on rectal examination. The cystoscopy revealed that the posterior urethra was slit-like, and a mass, about 3 cm, bulged to the bladder neck. The cystoscopy diagnosis was prostatic hyperplasia. Tumor markers, including prostate-specific antigen (PSA), carcinoembryonic antigen, and carbohydrate antigen 19.9, were within the normal range.

The imaging findings of patients are shown in

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figure 1. The ultrasound examination (figure 1A) showed a large, solid prostatic mass with a capsule and a well-defined border. Computed tomography (CT) showed that the mass was hypodense (23-47 Hu) with calcification in it (figure 1B). MRI revealed a large mass (10.1 × 6.9 × 9.7 cm^3) with a well-defined border involving the prostate. The mass was heterogeneously hyperintense on T1WI. The flaky hypointense signal was distributed in a slightly hyperintense background on T2-weighted imaging (T2WI). The mass was also hyperintense on both diffusion-weighted imaging (DWI) and ADC images. Dynamic contrast-enhanced MRI revealed that the mass had persistent progressive enhancement, and the time-signal intensity curve was of increasing type.

The patient underwent tumor resection. The pathological examination revealed a well-circumscribed lesion composed of loosely arranged spindle-shaped neural cells with myxoid stroma (figure 2A). The immunohistochemical analysis showed that the tumor cells were positive for Vim (figure 2B), S-100 (figure 2C), CD34 (figure 2D), MBP, and Ki-67 (+) ＜1%, and negative for SMA and desmin.

The patient was retrospectively examined for other manifestations of neurofibromatosis. His family and medical histories was normal. The physical examination did not show any café au lait spots or other cutaneous or mucosal stigmata that would have suggested neurofibromatosis. The CT scan of the patient was completely negative after the 3-month follow-up.

Table 1. Characteristics of prostate neurofibroma with NF-1 in published studies.

Reference	Age (year)	Symptoms	Treatment
Schoenberg et al. [19]	**4**	Urinary frequency, urgency, and nocturia	Suprapubic cystostomy tube
Blum *et al*. [20]	**31**	Prostate nodule on DRE	None (patient died)
Andrew *et al*. [3]	**35**	Prostate mass on DRE.	Observation
Manisha *et al*. [4]	**12**	Insidious onset of urinary obstruction	Surgical excision

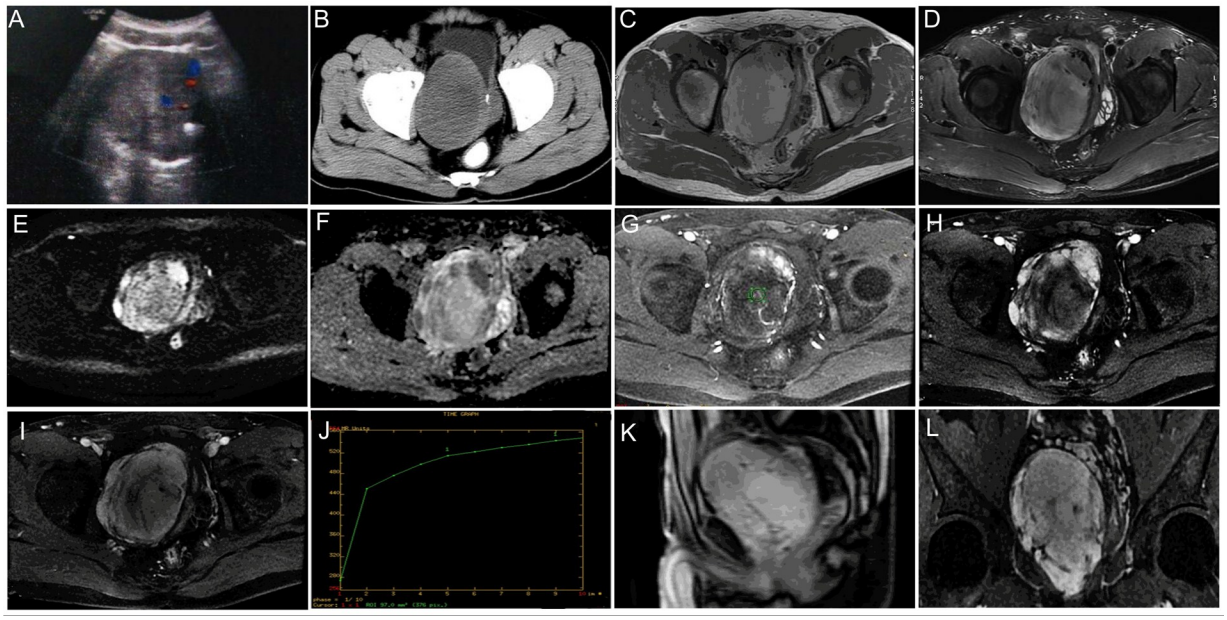


Figure 1. Imaging findings of patients. **(A)** Pelvic ultrasound revealed a hypoechoic irregular mass arising from the prostate. **(B)** CT showed that the lesion was hypodense with calcification in it. **(C)** MRI revealed a large heterogeneously hyperintense mass with a well-defined border involving the prostate on axial T1WI. **(D)** The mass was hyperintense (some part was hypointense compared with T1WI) on axial fat-saturated T2WI. **(E and F)** The mass was also hyperintense on DWI and ADC images. **(G)** Enhanced MRI showed that the lesion had a little enhancement on immediate post-contrast images. **(H and I)** The mass had a significant retention of contrast on 5- and 10-min-delayed images. (J) The time-signal intensity curve was of increasing type. **(K)** The sagittal T2WI showed that the mass was between the bladder and the rectum. **(L)** Coronal imaging of enhanced T1WI.

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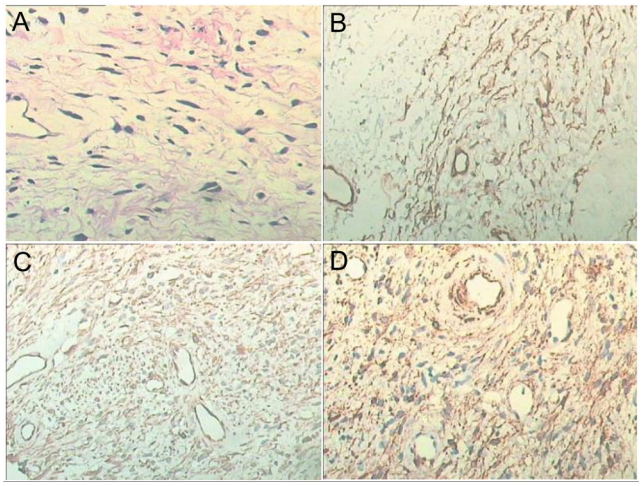


Figure 2. Pathological examination of patients. **(A)** Hematoxylin and eosin staining of prostate neurofibroma. **(B, C, and D)** Tumor cells were positive for Vim, S-100, and CD34.

DISCUSSION

NF1 is an inherited disorder that predominantly affects the skin and peripheral nervous system. It is caused by mutations in genes coding for neurofibromin located in the long arm of chromosome 17 (17q11.2). The diagnosis of NF-I is based on clinical criteria (table 2).

Neurofibroma is a benign tumor of the nerve sheath originating from the Schwann cells. It has three subtypes: localized, diffused, and plexiform. Localized neurofibromas are the most common type, accounting for about 90% of neurofibromas [5]. They are benign and slow growing; 60%–90% of patients do not have NF-1. Such neurofibromas are termed as solitary neurofibromas [6]. The diffused and plexiform neurofibromas are usually associated with NF-1. Neurofibromas associated with NF-1 may become malignant [7]. The investigation revealed no stigmata of NF-I. Therefore, the prostate mass of the patient was considered as a solitary neurofibroma without NF-1. Solitary neurofibromas have been reported to occur at the following locations: vertebra, retroperitoneal space, mandible, buccal mucosa, nose, bladder, spermatic cord, breast, common bile duct, abdominal wall, lower lip, oropharynx, and scrotum. However, no case report of prostate solitary neurofibroma is available in the literature.

Solitary neurofibromas most frequently affect young patients aged 20–30 years, with no sex predilection [8]. A solitary neurofibroma of the prostate may arise from the prostatic plexus [9]. Common clinical symptoms include recurrent urinary tract infection or urgency caused by tumor compressing the urogenital tract. PSA is generally within the normal range. These neurofibromas rarely undergo malignant transformation.

Image inspection is the main method for discovering and evaluating neurofibromas. Normally, solitary neurofibromas are round or oval, well-defined masses on imaging. On CT, neurofibromas are often hypodense masses with attenuation values of 20–30, which can result from a high content of fat or Schwann cells within the tumor. They may show heterogenous enhancement with attenuation values of 30–50 [10]. However, CT lacks the characteristics for definitive diagnosis.

On MRI, neurofibromas are usually heterogeneously hyperintense on T2WI and hypointense on T1WI. The “target sign” is a more specific sign of a neurogenic tumor, in particular, a neurofibroma [11]. The “target sign” on T2WI, which represents central low-intensity fibrosis surrounded by hyperintense myxoid stroma, can be observed in 55%–70% of neurofibromas [8]. Stroma enhancement is observed after contrast administration [10, 12]. Neurofibromas show no diffusion restriction on DWI and ADC images (if hyperintense in DWI may because of the shine-through effect) [13]. In addition, perfusion

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imaging may help differentiate benign neurofibromas from malignant lesions [14]. The characteristic MRI features are highly suggestive of the diagnosis. MRI is considered a valuable tool for diagnosing the disease. The imaging results of this study matched the findings in the literature, except that the mass was heterogeneously hyperintense on T1WI (might be related to steatosis and myxoma). The final diagnosis was based on the histopathological findings.

Differential diagnosis based on MRI findings includes the following diseases:

- Prostate cancer: Often occurring in the peripheral zone of the prostate, it is hypointense on T2WI and ADC images, hyperintense on DWI, and has "wash-out" enhancement on an MRI time–signal intensity curve. MRS shows a significant decline in citrate (CIT) and an increase in choline (CHO). PSA levels increase. In addition, the "T1 hemorrhage exclusion sign" is helpful to detect it on MRI [15].
- Benign prostatic hyperplasia: Occurring in the prostate transition zone, it involves spherical symmetrical expansion of the prostate, with uniform density, smooth edges, and no significant enhancement. MRS shows a significant increase in the concentration of CIT and increased level of CHO less than 15%.
- Prostatic solitary fibrous tumors: They are hypointense on T1WI and heterogeneous on T2WI, with multiple flow voids representing prominent vascular channels and marked or delayed enhancement on contrast T1WI. The latter enhancement feature is "geographical appearance" [16].
- Prostate leiomyoma: It is homogeneous and almost isointense on T1WI and T2WI, indicating pathological characteristics of abundant muscle or fibrous tissue. It shows homogeneous and obvious enhancement on contrast-enhanced imaging. It displays heterogeneous intensity when the tumor undergoes degeneration or necrosis.
- Prostate schwannoma: The imaging findings are similar to those of neurofibromas, but the heterogeneous intensity caused by cystic degeneration or necrosis is much more common in schwannomas than in neurofibromas [8].

Neurofibromas are not sensitive to radiation therapy or chemotherapy. They are usually treated with the surgical excision of the mass [10]. A frozen-section microscopic examination must be performed at the time of surgery to ascertain whether the tumor is benign or malignant. In the present case, the absence of mitotic activity, lack of necrosis, and infiltrative growth pattern suggested no malignancy. However, the treatment of choice for neurofibromas, in particular, for solitary ones, is still controversial. Some studies indicated that the best treatment for solitary neurofibromas was radical operation [17]. Others indicated that surgical resection was necessary only when the tumor caused pain or neurological deficiencies, and also when malignancy was strongly suspected [18]. Solitary neurofibromas are associated with a low local recurrence rate, if completely excised. The histopathological features of the patient in the present case indicated the presence of a benign tumor, which led to severe clinical symptoms. Therefore, the tumor was conservatively excised, and no signs of recurrence were noted on the CT scan after the 3-month follow-up.

In conclusion, this study reported the first case of a prostate neurofibroma without NF-1. Although prostate neurofibroma is a rare disease that may be difficult to diagnose, MRI has specific characteristics making it a valuable tool to diagnose the disease.

Table 2. Diagnosis of NF-I based on clinical criteria.

-Six or more café au lait spots, >0.5-cm diameter in the prepubertal age and >15-mm diameter in the postpubertal age.
-Axillary or inguinal freckles
-Optic glioma
-Two or more Lisch nodules: pigmented bilateral hamartomas, which appear as copular elevations on the iris surface -Distinctive bone lesion, sphenoidal dysplasia, dysplasia, or thinning of long cortical bones -First-degree relatives with NF-1
At least two of these clinical criteria should be met to diagnose NF-1

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