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By Dr. B. Shabarinath, Dr. Rahul Devraj, Dr. Megha S Uppin
& Dr. Nishika Madireddy

Nizam's Institute of Medical Sciences

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A Rare Case of Solitary Fibrous Tumor of Prostate - A Case Report

Dr. B. Shabarinath ^α, Dr. Rahul Devraj ^σ, Dr. Megha S Uppin ^ρ & Dr. Nishika Madireddy ^ω

Abstract- Solitary fibrous tumor (SFT), initially thought to be limited to the pleura can now be seen in numerous other sites. Prostatic solitary fibrous tumor is exceedingly rare and presents with nonspecific symptoms. We report a case of solitary fibrous tumor in a 43-year-old male patient who presented with obstructive urinary symptoms. A Trans rectal ultrasonography guided prostate biopsy was done. Microscopic examination revealed a lesion composed of spindle cell arranged in storiform pattern admixed with bands of collagen and haemangiopericytomatous vessels. Immunohistochemically the spindle cells stained positive for CD34 and STAT6 thus confirming the diagnosis of SFT. The patient underwent complete resection and the recovery was uneventful. Currently complete excision seems to be the most important prognostic factor. We also review the histologic mimics of SFT and the utility of STAT6 in aiding the diagnosis of SFT.

I. INTRODUCTION

Solitary fibrous tumor (SFT), an uncommon spindle cell tumor was first described in 1931 by Klemperer and Rabin.^{1,2} It accounts for less than 2% of all soft tissue neoplasms and was erroneously thought to be of mesothelial or submesothelial origin.²⁻⁵ However, advances in understanding the histogenesis and immunohistochemistry in the past few decades have shown that they arise from CD34 positive dendritic cells which are ubiquitously distributed throughout the

body.²⁻⁵ Previously thought to be limited to the pleural cavity and other mesothelial lined cavities it has now been established that SFTs can involve a variety of extrapleural sites like the breast, kidney, meninges, liver, bladder and seminal vesicle.⁵⁻⁷ However, SFT of the prostate is exceptionally rare and only a handful of cases have been reported in the literature. We report a case of solitary fibrous tumor of the prostate in a 43-year-old male patient presenting with obstructive lower urinary tract symptoms.

II. CASE REVIEW

A 43-year male patient presented to the Urology out patient with obstructive urinary symptoms such as a narrow stream, intermittency, straining for micturition with an International prostatic symptom score (IPSS) score of 19. His past history was insignificant and he had no other co morbidities. Ultrasonography revealed an enlarged prostate measuring 150 cc with the left lobe showing a heterogeneous echotexture and increased vascularity. The serum PSA levels were within normal limits (0.8 ng / ml) and a CECT revealed a heterogeneously enhancing mass lesion arising from the left lobe of prostate displacing the urethra to the right with loss of fat plane between the prostate and rectum. (Figure 1).

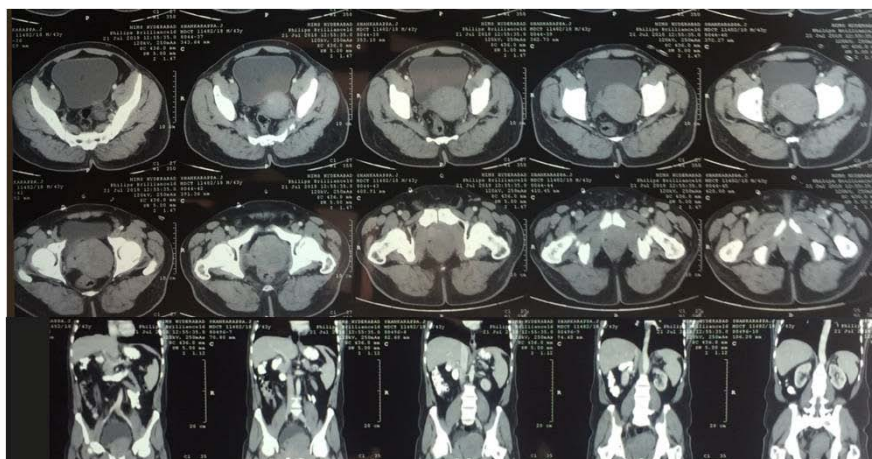


Fig. 1: CECT showing heterogeneously enhancing mass lesion arising from the left lobe of prostate displacing the urethra to the right with loss of fat plane between the prostate and rectum

Author ^α ^σ ^ρ ^ω: MCH Urology, Department of Urology, Nizam's Institute of Medical Sciences, Panjagutta, Hyderabad.
e-mails: shabarinathbijju@gmail.com, merahudevraj@rediffmail.com, megha_harke@yahoo.co.in, nishikareddy88@gmail.com

Trans-rectal ultrasonography guided biopsy from prostate was done showing features consistent with mesenchymal tumor. After complete surgical profile patient was posted for open prostatectomy.

Intra-operatively left lobe of prostate gland was enlarged, complete gland removal was done and specimen was sent for histopathological examination.

Gross examination revealed complete enlargement of the prostate measuring 20 x 11 x 3.5 cm with nodular external surface. The cut surface had whitish firm appearance without any areas of necrosis or

haemorrhage. (Figure 2) Microscopic examination revealed a lesion composed of spindle cells arranged in a storiform pattern with intervening bundles of collagen. Numerous staghorn vessels were observed within the lesion. No necrosis, atypia or increased mitotic activity was noticed. Immunohistochemically the spindle cells were positive for STAT6 and CD34. (Figure 3).



Fig. 2: Gross prostatectomy specimen showing enlarged prostate with nodular appearance

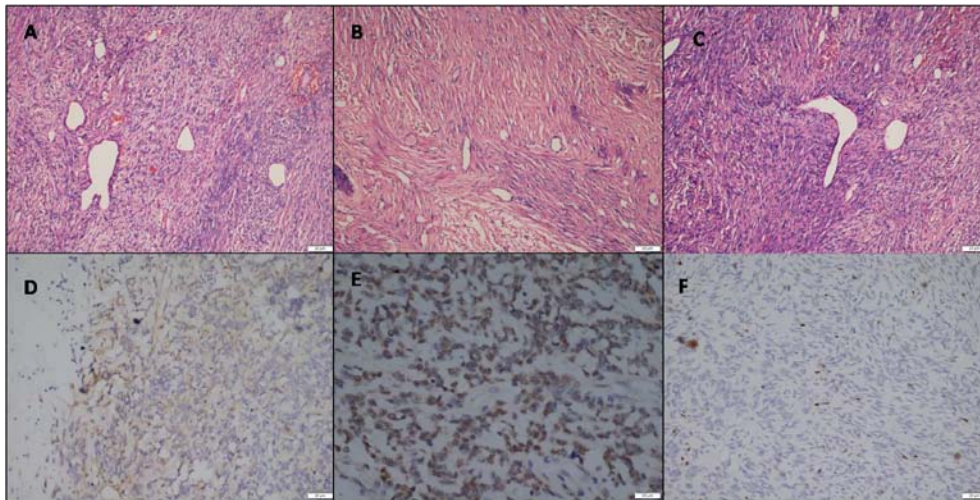


Fig. 3 : A, B, C the microscopic sections showing spindle cells arranged in patternless sheets separated by collagen bundles and classic staghorn vasculature. (D) Immunohistochemical positivity for CD34, (E) STAT6 and (F) low Ki67 index

Based on the above H&E and immunohistochemical findings we arrived at a diagnosis of a Solitary Fibrous tumor of the prostate.

Postoperatively patient recovered well with improvement in IPSS score to 8. Patient is in close follow up.

III. DISCUSSION

Mesenchymal lesions of the prostate are rare neoplasms and often pose a diagnostic challenge due to an overlap in their histomorphological as well as

immunohistochemical features.⁷⁻⁹ Mesenchymal lesions of prostate encompass a wide range of lesions including stromal proliferation, solitary fibrous tumour, leiomyoma, Inflammatory myofibroblastic tumor, Schwannoma, liposarcoma, angiosarcoma, and rhabdomyosarcoma. While prostatic stromal proliferation is the most common mesenchymal neoplasms in the prostate, solitary fibrous tumors are regarded as the least common.⁷⁻⁹

Solitary fibrous tumors of the prostate are usually seen in middle-aged adults ranging between 50-87 years with a mean age of presentation of 55 years

and present as slow-growing masses.¹⁰⁻¹² The tumor may remain asymptomatic initially however with increase in size they most commonly present with urinary retention, increased frequency, urgency, dysuria, hematuria, constipation and groin and abdominal pain.^{10,11} The PSA levels are usually within normal limits and hypoglycemia reported in pleural solitary fibrous tumors is not frequently observed in prostatic solitary fibrous tumors.^{12,13} The tumor size shows a wide size distribution ranging from 2-14cm and can weigh anywhere between 5 gm to 170 g.¹¹⁻¹³ In fact, in a case reported by Gharaee-Khermani et al¹¹ the prostate weighed more than 700 gm.

Gross examination of SFTs usually reveals a well circumscribed grey white mass whose cut surface is nodular and may show a whorling pattern. Histomorphologically it is composed of spindle cells arranged in a herringbone or storiform pattern or the so-called patternless sheets comprised of alternating hyper and hypocellular areas or a combination of all the above patterns admixed with thick bands of collagen fibers and intervening haemangiopericytomatous or staghorn vessels.¹³⁻¹⁵ Solitary fibrous tumors are usually benign however increased cellularity, high mitotic figures (>4/10 HPF), nuclear pleomorphism, the presence of necrosis and infiltrative borders may favor the diagnosis of malignant solitary fibrous tumor^{10,13} However these criteria are not definite and don't seem to have any significant impact on the outcome.^{10,13} Immunohistochemically the cells are positive for CD34, Vimentin, and bcl2, CD99, and SMA but negative for S100, Actin, desmin, epithelial markers and CD117.¹⁰⁻¹⁹ These lesions may be associated with adjacent prostatic adenocarcinoma. Two cases, one reported by Moureau-Zabotto et al¹² and one reported in a case series by Herawi et al¹⁹ additionally showed adenocarcinoma in the adjacent prostatic parenchyma.

The differential diagnosis for these tumors can be other mesenchymal lesions of the prostate. Stromal nodules and stromal tumor of uncertain malignant potential (STUMP) are both composed of spindle cells intermixed with blood vessels but show entrapped normal prostatic glands which are usually absent in SFTs. STUMP also shows a positivity for CD34, however, it is also positive for PR whereas SFTs are negative for PR.¹³ The other tumors of mesenchymal origin include leiomyoma, leiomyosarcoma, rhabdomyosarcoma, Inflammatory myofibroblastic tumor, fibrosarcoma, synovial sarcoma and GIST.^{13,17,18} In contrast to SFT leiomyoma and leiomyosarcomas show a more uniform fascicular pattern and are positive for desmin and SMA.^{13,19} Rhabdomyosarcomas are usually encountered in the younger age group with embryonal morphology and positivity for myogenin and desmin.¹³ Fibrosarcoma and synovial sarcoma can be differentiated from SFT by their bimodal pattern and negativity for CD34.^{13,14} IMFT usually shows inflammatory

cells admixed with spindle cells and positivity for ALK.^{13,18-20} GIST which also shows spindle cell morphology can be differentiated from SFT with the help of CD117 which is positive in GIST but negative in SFT.¹³⁻¹⁹ Sarcomatoid carcinoma is another important tumor which needs to be differentiated from SFT and this is done with the aid of keratins which are positive in Sarcomatoid carcinomas^{13, 18}

Classically the immunohistochemical panel of CD34, CD99, and bcl-2 has aided the diagnosis of SFT, however, it has now been proven that these markers have a poor specificity. CD34 is positive in other spindle cell lesions like GIST, stromal tumors of prostate and Schwannoma thus making it difficult to differentiate them from SFT.^{17,21-24} Recent genomic studies have identified a fusion gene NAB2-STAT6 in almost 100% of the SFTs.^{11,24-26} This has led to the development of a novel nuclear marker STAT6 which is considered a highly specific and sensitive marker for SFT. NAB2 is located in the nucleus and encodes zinc finger transcription factors involved in regulating differentiation and proliferation. STAT6, a cytoplasmic protein relocates to the nucleus on activation and is involved in signal transduction.^{22,24} The NAB2-STAT6 fusion gene exerts an oncogenic effect by induction of EGR mediated transcription.²⁴ Except for few ambiguous meningeal lesion showing features of both meningioma and SFT, NAB2-STAT6 fusion gene has been seen almost exclusively in SFT.²⁴⁻²⁶ Thus, STAT6 nuclear expression is now identified as the most reliable marker of SFT.²¹⁻²⁴ ALDH1, a cytoplasmic protein has been identified in 75% of the SFTs and a study by Guner et al²² has shown that a combination of ALDH1 and STAT6 greatly increased the specificity.

Extrapleural SFTs are generally thought to have an indolent course in comparison to their pleural counterparts however a local recurrence or distant metastasis has been noticed in upto 20% of the cases.^{10,12,21,24} Solitary fibrous tumors are relatively unresponsive to chemotherapy or radiotherapy, thus irrespective of the benign or malignant nature of the tumor a complete excision of the tumor with negative margins is the treatment of choice and is also the most important prognostic factor.^{12,15-18}

IV. CONCLUSION

We have reported a rare case of Solitary fibrous tumor arising in the prostate in a 43-year-old male patient presenting with obstructive urinary symptoms. The lesion comprised of haphazardly arranged spindle cells admixed with bands of collagen and stag horn blood vessels. In concordance with recent studies, our case was showed a nuclear positivity for CD34 and STAT6. The patient underwent complete excision and had an uneventful recovery. Prostatic SFTs are extremely and it is extremely important to differentiate

them the from other spindle cell lesions. Owing to the rarity of prostatic solitary fibrous tumor, there is limited data regarding the clinical behaviour as well as the outcome of the tumor, thus, it is important to identify the lesion and keep the patient on close follow-up.

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