Oncology

Solitary fibrous tumor behind the pubic bone: A rare case report

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A B S T R A C T
Solitary fibrous tumors (SFTs) are a rare type of soft tissue tumors and there have been only a few reported cases of SFTs at the perineum region. We report a SFT below the pubic bone in a 35-year-old male causing compression on his corpus spongiosum. The patient underwent en bloc tumor resection with preservation of spermatic vessels and the bladder neck. Pathological evaluation and immunohistochemistry (IHC) staining with CD34( - ), CD99 ( ), STAT6( - ), S100( - ) and Desmin( - ) confirmed the diagnosis of SFT. Surgery plays a key role in treatment strategies and pathological examination with IHC is important in the diagnosis of SFTs.

Introduction

Solitary fibrous tumor (SFT) is a rare condition which accounted for less than two percent of all soft tissue tumors. 1 SFT can arise at different sites in the body, most commonly occurred in the thoracic and peritoneal cavities, followed by extremities, and head and neck region. SFTs arising from soft tissue of perineum are extremely rare. Among different sites of tumor, there are certain significant differences in clinical presentation and management of SFTs.

Surgery is considered as the primary treatment of SFTs, but chemotherapy and radiation therapy could also be indicated in some cases. 1 However, there have not been any standard treatment strategies for these tumors. Therefore, information from clinical cases could be useful for clinicians to define better treatment approach for tumors at each site. In this paper, we present a case with SFT locating at the base of the penis which might be one of the first cases of SFTs at this site.

Case presentation

A 35-year-old male, with no past medical histories, came to our institution after noticing a mass in his left inguinal region which was painless and gradually increased in size for five months. The mass rapidly enlarged in the last two months and the patient experienced increasing urinary frequency and urgency as well as erectile dysfunction. On examination, there was a hard and tender, about 10-cm mass located in the soft tissue around pubic bone, without skin involvement.

On digital rectal exam, the mass lied outside of rectal lumen. No enlarged inguinal lymph nodes and testicular abnormalities were detected.

Ultrasound showed a mass of mixed echogenicity at the base of the penis and scrotum which caused bilateral spermatic cord compression but did not extend into abdominal cavity. On magnetic resonance imaging, there was a large mass with an anterior-posterior x width x cranio-caudal diameter of 100 73 mm located in soft tissue anterior and superior to the base of the penis and below the pubic bone, causing compression on the root of corpus spongiosum. It had irregular borders but demonstrated no fatty infiltration.

The patient then underwent operation for tumor resection. During surgery, the tumor was found to surround the base of the penis and extend posteriorly to the anterior surface of the prostate but did not invade surrounding tissues (Fig. 1). En-bloc tumor resection was performed carefully, with preservation of spermatic vessels and the neck of the bladder (Fig. 1).

Pathological specimen of the tumor showed spindle cells with multiple oval nuclei, smooth nuclear chromatin and eosinophilic cytoplasm, with alteration of poorly cellular and more cellular areas and large branching thin-walled vessels (Fig. 2). Besides, there were no malignant features, including high cellularity, mitosis, as well as necrosis. Immunohistochemistry test results were positive with CD34, CD99, STAT6 (in which STAT6 positivity was confined to the nucleus) and negative with CK, SMA, S100 and Desmin (Fig. 3). The final diagnosis was therefore solitary fibrous tumor. Follow-up examination and imaging assessment

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Discussion

Solitary fibrous tumors (SFTs) were first recognized as a distinctive entity by Klempere and Rabin in 1931. Most SFTs located in thoracic cavity (30%, including pleura, lungs and mediastinum), the peritoneal cavity, retroperitoneal space and pelvis (30%), and the head and neck region (about 20%). Among the remaining sites, SFTs behind the pubic bone are extremely rare. To our best knowledge, there have not been any reports of SFT at this site.

In a case-series of 99 patients with SFTs, most extrathoracic cases presented as a slow-growing, painless mass or further symptoms attributable to mass effect such as abdominal pain, urinary retention, dysuria and tenesmus in cases with perineal SFT. Compared to pleural forms, extrapleural SFTs tend to have more aggressive clinical course and biological behavior, which might related to different rearrangement pattern of NAB2/STAT6 fusion gene due to paracentric inversion on chromosome 12q13.

In our patient, the differential diagnosis might include mesotheliomas and sarcomas such as gastrointestinal stromal tumors (GIST), synovial sarcoma, liposarcoma and fibromyxoid sarcoma. Especially, in case of SFTs involving perineum, the tumor also has to be differentiated from well-differentiated liposarcoma and prostatic stromal tumor. These disorders shared certain histopathological features on HE-stained specimens that can lead to diagnosis difficulties. Thus, IHC has an important role in determining a tumor as SFTs. Markers that are commonly used to diagnose SFTs include CD34, CD99, STAT 6, S100 and Desmin. In which, CD34 has a sensitivity of 95%–100% and strong nuclear expression of the C-terminal part of STAT6 has a sensitivity of 98% and a specificity of more than 85%. In this patient, the clear border between the tumor and the prostate as well as STAT6 staining positivity made the diagnosis of prostatic stromal tumor unlikely. Besides, although some well differentiated liposarcoma may be positive for STAT6, the stain positivity was confined to the nucleus (Fig. 3), which favored the diagnosis of SFT rather than well-differentiated liposarcoma.

Treatment strategies of SFT might include surgery, chemotherapy and radiation therapy. However, in cases with local-regional disease, chemotherapy and radiation therapy is hardly indicated, unless the tumor is unresectable or negative margin cannot be achieved. As a result, surgery plays the most important role, in which en bloc resection could guarantee a curative therapy. In our case, the patient has a large tumor located behind the pubic bone. Resection of this tumor can potentially cause damage to surrounding structures such as the bladder, the prostate, spermatic vessels and dorsal penile vessels. Careful dissection of these anatomic structures is necessary to avoid post-operation complications, including bladder fistula, sexual dysfunction and testicular atrophy.

Conclusion

In conclusion, diagnosis of SFTs can be challenging, in which pathological examination with IHC plays an essential role. Surgery to completely resect the tumor is the first choice in the treatment approach while chemotherapy and radiation therapy have limited contribution.

Author contributions

Hau X. Nguyen: Surgeon, revised manuscript. Hung V. Nguyen: Assistant surgeon, wrote manuscript. Long T. Nguyen: Assistant surgeon, wrote manuscript. Luc T. Dao: Main surgeon, follow up the patient.

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None.

Consent

The publication of this study has been consented by the relevant patient.

Ethical approval

The study was approved by our research committee, Hanoi Medical University Hospital, Hanoi, Vietnam.

Guarantor

Hau X. Nguyen, M.D, Ph.D.

Declaration of competing interest

None.
Fig. 2. Histological characteristics in low power (A and B) and high power (C and D).
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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2020.101127.

Fig. 3. Pathology and immunohistochemistry staining results with HE stain (A), positive with CD34 (B), CD99 (C), STAT6 (confined to the nucleus) (G) and negative with CK (D), Desmin (E), S100 (F) and SMA (H). The final diagnosis was therefore solitary fibrous tumor.
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