Int J Clin Exp Pathol 2019;12(1):212-216
www.ijcep.com /ISSN:1936-2625/IJCEP0085495

Original Article

Lipomatous hemangiopericytoma of the corpus spongiosum: a case report and review of the literature

Chuang Liu*, De-Gang Ding*, Lei Shan, Xiao-Yu Duan, Tong-Yu Ji, Rui-Jin Zhou, Tao Du

*Department of Urology, Henan Provincial People's Hospital, Zhengzhou, China. *Equal contributors.*

Received September 14, 2018; Accepted September 25, 2018; Epub January 1, 2019; Published January 15, 2019

Abstract: Background: Hemangiopericytoma (HPC) is an uncommon soft tissue tumor arising from pericytes. The urogenital system is rarely affected. Methods: The review of the literature used the PubMed database which was searched up to March 2015. Results: Herein, we report the first case of lipomatous HPC of the corpus spongiosum in a 37-year-old man in China. The lesion presented as a quickly growing mass. Contrast enhanced CT showed a heterogeneous fatty mass with a multifocal enhancing soft-tissue component. Microscopically, the neoplasm was composed of spindle cells, a mature fat component and collagenous stroma. The mitotic index was low at 1 to 3 mitoses per 10 high-power fields. Immunohistochemically, STAT6, Bcl-2, CD99 and CDK4 were positive; CD34 and SMA were negative. The mature adipocytes were positive for S-100. Ki-67 expression was found in approximately 5% of the tumor cells. Surprisingly, there was a diffuse and strong nuclear expression of MDM2, but, no amplification of MDM2 was demonstrated by FISH. An adequate excision of the tumor was performed. Conclusion: No local recurrence or distant metastases occurred during the 18-month follow-up. In view of the unpredictable biological behavior of this tumor, a long follow-up period is mandatory.

Keywords: Hemangiopericytoma, corpus spongiosum, S-100, MDM2

Introduction

Hemangiopericytoma (HPC) is an uncommon soft-tissue vascular tumor arising from pericytes, which was first described by Stout and Murray in 1942 [1]. Nowadays, HPCs have been re-classified under the umbrella 'extrapleural solitary fibrous tumour (SFT)' by the new World Health Organization (WHO) classification of soft-tissue tumors [2]. SFTs are characterized by their histologic appearance, but they have a wide spectrum of morphologic variation, as well as of biological behavior [3]. The fat-forming variant of SFT (lipomatous HPC) and SFT share similar clinical, histologic and immunohistochemical features, except for the presence of mature adipocytes [3]. Lipomatous HPC has been described in several anatomical sites, but the lower extremities and the retroperitoneum are the most commonly affected sites [4, 5]. Long-term follow-up is recommended, even after successful management [6]. The purpose of this article is to report that this tumor occurs in the corpus spongiosum, the first such report.

Case report

A 37-year-old man presented to our outpatient clinic whose chief complaint was of a quick-growing mass in his penile shaft for about three months. He complained of some pain during sexual intercourse. To some extent, erectile function was affected. Urethral discharge, gross hematuria, fever, and any voiding symptoms were absent. There was no history of surgery, injury, or drug abuse. On physical examination, a firm and fixed mass with a mild tenderness was located on the bottom of the penile shaft. There was no enlargement of the inguinal lymph nodes. A complete blood cell count, serum biochemistry tests, urinalysis, and serum tumor markers indicated no abnormalities. Through rigid urethroscopy, we found that the urethral lumen was intact. A contrast-enhanced penile CT revealed a 57-mm diameter solid neoplasm arising from the corpus spongiosum (**Figure 1**). The tumor displayed a heterogeneous soft tissue density (**Figure 1**). Importantly, a lot of dense fatty tissue was

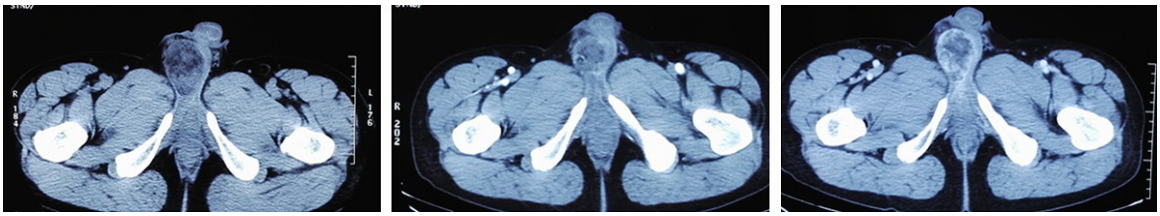

Figure 1. Contrast enhanced penile CT demonstrating a 57-mm diameter solid neoplasm arising from the corpus spongiosum. The mass was well circumscribed and was largely composed of fat and a remarkably sized, well-enhancing soft-tissue component.

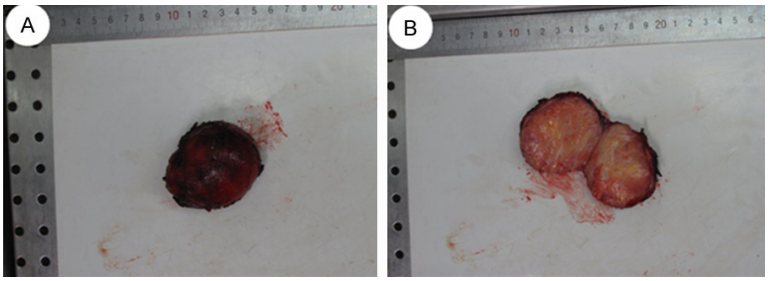

Figure 2. The tumor is brown in color, void-shaped and 6 cm in diameter.

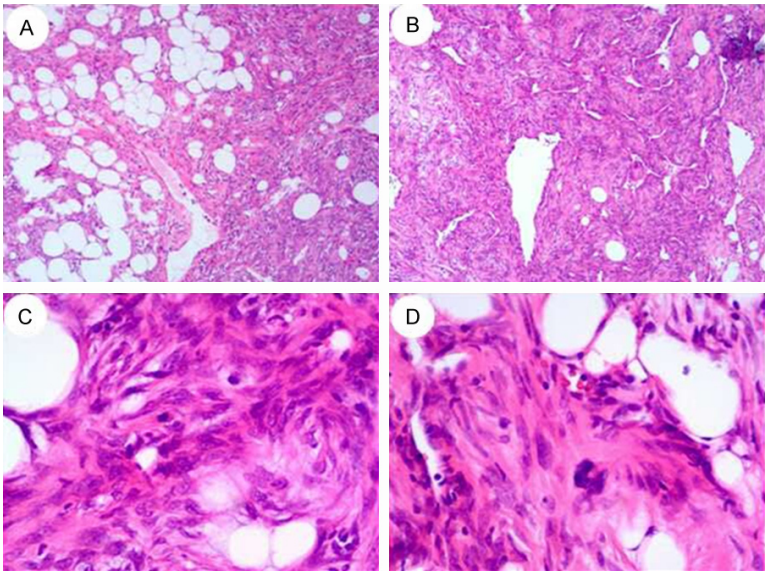

Figure 3. Histopathological examination of the mass reveals that the tumor is composed of hemangiopericytoma-like areas admixed with areas of mature adipocytes.

detected in the lesion (**Figure 1**). The whole mass was surgically removed. The adjacent organs such as the right testis and the corpora spongiosum remained intact. The biopsies of the remaining corpus cavernosum indicated an adequate resection margin. Grossly, a solid ovoid-shaped tumor (6 cm in diameter) was well demarcated and seemed to be encapsulated (**Figure 2**). On its surface, there was some venous engorgement. Under the microscope, the tumor consisted of spindle cells with 1 to 3 mitoses per 10 high-power fields (HPF), a mature fat component, collagenous stroma and branching vessels (**Figure 3**). The immunochemistry was as follows: STAT6 (+), Bcl-2 (+), MDM2 (+), CD68 (+), CDK4 (+), CD99 (+), ER (+), PR (+), S-100 (+), Ki-67 (+), CD34 (-), SMA (-) (**Figure 4**). The mature adipocytes were positive for S-100. Ki-67 expression was found in approximately 5% of the tumor cells. A fluorescence in situ hybridization (FISH) for MDM2 was also performed on paraffin-embedded tissue sections. No amplification of MDM2 was demonstrated by FISH (**Figure 5**). One and a half years after the surgery, no sign of local recurrence or distant metastases were identified on the chest, abdomen, or the penis on CT (**Figure 1**). However, the patient was afflicted with severe erectile dysfunction (ED). Phosphodiesterase-5 (PDE-5) inhibitors provided no improvement in erectile function.

Discussion

Lipomatous HPC possesses a peak incidence in middle-aged adults and has a slightly increased propensity to occur in males [4]. Most tumors tend to present as longstanding painless masses. Slowly growing lesions may cause symptoms due to their pressure effects on adjacent structures. Radiologically, they are

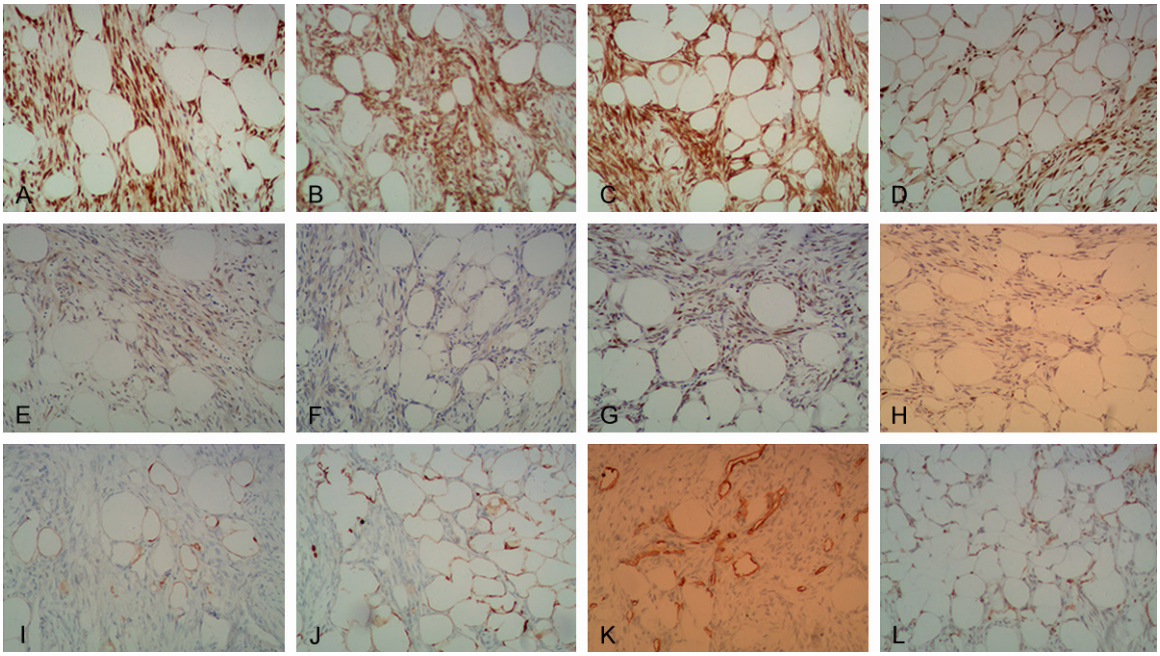

Figure 4. Immunohistochemistry indicated a strong positivity for STAT6 (A), Bcl-2 (B), CD68 (C) and MDM2 (D). Focal weak expressions of CDK4 (E), CD99 (F), ER (G) and PR (H), were also found in this tumor. The mature adipocytes were positive for S-100 (I). Ki-67 (J) expression was found in approximately 5% of the tumor cells. CD34 (K) and SMA (L) were negatively expressed in the tumor cells.

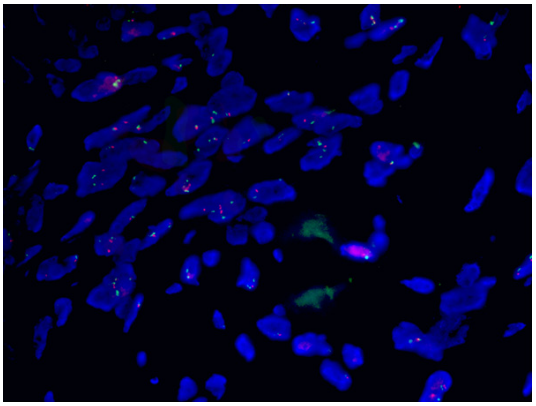

Figure 5. No amplification of MDM2 was demonstrated by FISH. GSP MDM2 (red)/CSP12 (green) = 1.2<2.2.

large well-circumscribed solid masses, with prominent feeding vessels and fatty components [5].

Tumors with typical histologic features usually behave in an indolent manner. The neoplasm was composed of variable mixtures of spindle cells with collagenous stroma, mature adipose tissue and branching hemangiopericytic vessels [4]. The mitotic index was low at 1 to 3 mitoses per 10 HPF [4]. Malignant lipomatous HPC is rarely encountered. Most tumors have a mitotic index of >4/10 HPF and show at least focal hypercellularity and moderate atypia and over half have areas of necrosis [7]. In addition to mature adipose tissue, tumors may contain multi-vacuolated lipoblasts as well as some areas resembling atypical lipomatous tumor (ALT) [7]. The two latter features seem to be much more common in the malignant subset of lipomatous HPC [7]. The neoplasms exhibiting malignant histologic features have potential for aggressive behavior. Immunohistochemistry contributes to the diagnosis of SFT. CD34, Bcl-2 and STAT6 are considered to be key positive markers in SFT [8]. As its fat-forming variant, lipomatous HPC typically shows diffuse and strong positivity for vimentin, CD99 and CD34 [4]. About half of the tumors show reactivity for Bcl-2 [4]. Some cases can display variable focal epithelial membrane antigens, such as S-100 and SMA expression [4]. However, lipomatous HPC is usually negative for CD31, desmin and cytokeratins [4].

STAT6 immunohistochemistry is a valuable tool for the distinction of SFT from its mimics, especially in the setting of CD34-negative SFT or its fat-forming variant [9, 10]. STAT6 is amplified in

a subset of dedifferentiated liposarcoma, leading to diagnostic confusion with SFT [11]. The degree of STAT6 expression in SFT is generally more diffuse and of a stronger intensity than in DDL [11, 12]. Moreover, STAT6 expression in SFT or its fat-forming variant is predominantly nuclear, while DDL expresses this marker in both nuclear and cytoplasmic components [10-12]. Lipomatous HPC may also resemble well-differentiated liposarcoma (WDL) but lacks the atypia seen in the adipocytes of WDL [3]. The vast majority of WDL and DDL cases show nuclear expressions of CDK4 and MDM2, which are typically absent in lipomatous HPC [3]. Another useful observation is that lipomatous HPC diffusely expresses CD99 which is not generally seen in WDL [7, 13].

The prognosis is usually excellent for the majority of lipomatous HPC cases with classical histological features, but the neoplasms with malignant features are predictive of poorer outcomes [7]. Since these are clinically aggressive, lipomatous HPC cannot be reliably distinguished from those that will behave indolently, so it is crucial that patients are followed up on a long-term basis [7]. Surgical resection remains the gold standard for the treatment for localized diseases. The emphasis is on obtaining negative margins as any residual microscopic lesion negatively affects survival. At this time, there is no robust evidence for the routine use of preoperative or postoperative radiation therapy or adjuvant chemotherapy.

In our case, a 37-year-old man presented with a quick-growing mass in his penile shaft for about three months. Erectile function was affected due to this painful mass. A contrast-enhanced penile CT also showed a large heterogeneous mass which was largely composed of fat and a well-enhancing soft-tissue component. These radiologic features and the property of tumor growth indicated that the tumor could be aggressive. However, the neoplasm showed the typical histologic appearance of lipomatous HPC. The mitotic index and the Ki-67 expression were both low in tumor cells. Furthermore, this tumor lacked the atypia, lipoblast and necrosis usually seen in malignant lipomatous HPC, DDL or WDL. Immunohistochemistry indicated a strong positivity for Bcl-2. Focal weak expressions of CD99 and CDK4 were also found in this tumor while CD34 expression was negative. Importantly, there was a diffuse and strong nuclear expression of STAT6. To our surprise, tumor cells showed a strong nuclear expression of MDM2 which is typically positive in DDL or WDL but is negative in classical lipomatous HPC. The conflicting results led us to carry out FISH for MDM2, which ruled out the amplification of MDM2. Based on all these features, we determined that our case should be regarded as lipomatous HPC. The whole mass was adequately removed because the biopsies of the remaining corpus cavernosum indicated the margins were negative.

Conclusion

The lipomatous HPC was completely removed. There was no local recurrence or metastases during the 18-month follow-up. However, in view of the low aggressive potential of this tumor, we monitor the patient regularly.

Disclosure of conflict of interest

None.

Address correspondence to: Rui-Jin Zhou and Tao Du, Department of Urology, Henan Provincial People's Hospital, 7 Weiwu Road, Zhengzhou 450003, China. Tel: +86-371-65580608; Fax: +86-371-65580716; E-mail: zrjin_ur@outlook.com (RJZ); lcg78032@163.com (TD)

References

[1] Stout AP and Murray MR. Hemangiopericytoma: a vascular tumor featuring zimmermann's pericytes. Ann Surg 1942; 116: 26-33.

[2] Fletcher CD. The evolving classification of soft tissue tumours - an update based on the new 2013 WHO classification. Histopathology 2014; 64: 2-11.

[3] Thway K, Ng W, Noujaim J, Jones RL and Fisher C. The current status of solitary fibrous tumor: diagnostic features, variants, and genetics. Int J Surg Pathol 2016; 24: 281-292.

[4] Guillou L, Gebhard S and Coindre JM. Lipomatous hemangiopericytoma: a fat-containing variant of solitary fibrous tumor? Clinicopathologic, immunohistochemical, and ultrastructural analysis of a series in favor of a unifying concept. Hum Pathol 2000; 31: 1108-1115.

[5] Park CY, Rho JY, Yoo SM and Jung HK. Fat-forming variant of solitary fibrous tumour of the pleura: CT findings. Br J Radiol 2011; 84: e203-205.

[6] Jing HB, Meng QD and Tai YH. Lipomatous hemangiopericytoma of the stomach: a case report and a review of literature. World J Gastroenterol 2011; 17: 4835-4838.
[7] Lee JC and Fletcher CD. Malignant fat-forming solitary fibrous tumor (so-called "lipomatous hemangiopericytoma"): clinicopathologic analysis of 14 cases. Am J Surg Pathol 2011; 35: 1177-1185.
[8] Geramizadeh B, Marzban M and Churg A. Role of immunohistochemistry in the diagnosis of solitary fibrous tumor, a review. Iran J Pathol 2016; 11: 195-203.
[9] Yoshida A, Tsuta K, Ohno M, Yoshida M, Narita Y, Kawai A, Asamura H and Kushima R. STAT6 immunohistochemistry is helpful in the diagnosis of solitary fibrous tumors. Am J Surg Pathol 2014; 38: 552-559.
[10] Creytens D and Ferdinande L. Diagnostic utility of STAT6 immunohistochemistry in the diagnosis of fat-forming solitary fibrous tumors. Appl Immunohistochem Mol Morphol 2016; 24: e12-13.
[11] Doyle LA, Tao D and Marino-Enriquez A. STAT6 is amplified in a subset of dedifferentiated liposarcoma. Mod Pathol 2014; 27: 1231-1237.
[12] Demicco EG, Harms PW, Patel RM, Smith SC, Ingram D, Torres K, Carskadon SL, Camelo-Piragua S, McHugh JB, Siddiqui J, Palanisamy N, Lucas DR, Lazar AJ and Wang WL. Extensive survey of STAT6 expression in a large series of mesenchymal tumors. Am J Clin Pathol 2015; 143: 672-682.
[13] Thway K, Flora R, Shah C, Olmos D and Fisher C. Diagnostic utility of p16, CDK4, and MDM2 as an immunohistochemical panel in distinguishing well-differentiated and dedifferentiated liposarcomas from other adipocytic tumors. Am J Surg Pathol 2012; 36: 462-469.