Letters to the Editor

An infrequent plexiform variant of schwannoma of the glans penis: a rare finding

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Dear Editor,

I am Dr Tzu-Chun Lin, from the Department of Dermatology, China Medical University Hospital, Taichung, Taiwan, China. We write to you to present a rare case of plexiform variant of schwannoma on the glans penis.

Schwannomas, also named neurilemmomas, of the penis are extremely rare even though the innervation of the genital region is exuberant and complex. There have been 27 penile schwannomas published in the literature [1], among which only four cases were present on the glans. However, to our knowledge, no plexiform type of schwannoma involving the glans has been previously mentioned.

We report on a 44-year-old man who presented with an ovoid-shaped, elastic nodule on the dorsum of the glans penis (Figure 1). He first noticed a new growth on the penis 1 year earlier. The mass was skin-colored, asymptomatic and grew slowly, remaining constant in size. He denied any previous history of sexually transmitted diseases, penile trauma, auto-injection of enhancing agents or abnormal sexual behavior. Physical examination revealed a semitransparent, elastic nodule, measuring up to 1.1 cm in greatest diameter, protruding from the dorsal aspect of the glans penis. He underwent simple total resection of the tumor with adequate surgical margins. Postoperatively, his erectile function was normal.

Histologically, in the submucosal tissue, there are several nodular and lobulated lesions with incomplete circular structures that are surrounded by a thin and stretched perineurium (Figure 2A). Nodular and lobulated lesions are composed of a combination of a predominantly cellular zone of Antoni A areas characterized by highly cellular spindle cells with

Figure 1. A symptomless, ovoid, elastic exophytic lesion arising from the dorsal corona of the glans penis of 1-year duration.
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nuclear palisading, and an interspersed loose cellular zone of Antoni B areas consisting of sparse spindle cells in myxoid stroma with focal microcystic change. (Figure 2B). Under immunohistochemical study, the tumor cells disclosed diffuse and intense staining for S-100 protein (Figure 2C), supporting the neural origin of the tumor cells, whereas malignant schwannomas are usually characterized by very faint and focal S-100 protein expression. The cells showing positive immunohistochemical staining for epithelial membrane antigen were uniquely present within the capsule (Figure 2D). Based on clinical presentation, histological features and immunohistochemical studies, the nodule was confirmed to be a benign plexiform schwannoma.

Plexiform schwannomas mostly affect younger adults, with no pronounced sex predilection. Most plexiform schwannomas are solitary, sporadically occurring tumors. Plexiform schwannomas typically arise from superficial tissues (79%), occurring most commonly in the head and neck region (23%) and in 15% of cutaneous schwannomas [2]. In the male genital neoplasms, there have been two cases of benign intratesticular schwannoma reported in the published literature, including one report of an unusual plexiform variant growth of testicular schwannoma [3]. Embryologically, both the clitoris and glans penis originate from the genital tubercle with structural differentiation. Five cases of schwannoma of the clitoris have been published in the literature, among which two cases belonged to the plexiform subtype, including a congenital plexiform schwannoma of the clitoris [4, 5]. Herein, we report a case of plexiform schwannoma on the glans penis. Although the embryologic origins of the male glans and female clitoris are identical, plexiform schwannoma has not yet been mentioned on the glans.

Schwannomas are encapsulated lesions that are usually eccentrically attached to nerves and easily shelled out without any appreciable nerve insult. The high frequency of intratumoral axons in conventional and cellular schwannomas contradicts the conventional paradigm that the pathogenesis of these lesions is a nearly pure population composed solely of Schwann cells or showing Schwannian differentiation. Surprisingly, this raises the possibility that plexiform schwannoma or other variants may represent biologically distinct entities, suggesting a diversified pathway of tumorigenesis for varying types of all schwannomas [6].

In one cytogenetic study of 14 sporadic schwannomas [7], the predominant genetic aberrations were chromosomal imbalances in 79% of cases, among which the number of gene losses was higher than gains. The present analysis of schwannomas revealed loss of chromosome 22 material, which is identified to be a tumor suppressor with loss of its protein product merlin as the most prominent alteration expected in slightly more than half of the cases, included loss of 22q material through monosomy 22 or unbalanced structural rearrangements, followed by loss of a sex chromosome and trisomy 7. The limited number of case studies available shows that the cytogenetic features of schwannomas are not dependent on the site of origin [8]. Multiple schwannomas are seen in association with neurofibromatosis type 2 and schwannomatosis, with both having their basis in mutations in NF-2 gene (on chromosome 22q12) [9]. Patients with neurofibromatosis type 2 have germline mutations in one allele. Now it has been clearly elucidated that before the development of nerve sheath tumors a
second deletion or mutation of the other allele in NF-2 gene is required. In other words, both copies of the gene have a loss of function. However, these studies do not identify the precise role of the different genetic mutations in determining the conformational change of cytoskeleton and cellular composition.

Plexiform schwannomas are benign tumors. The rate of local recurrence is extremely low after complete resection with free surgical margins [10]. Recurrence may happen when excised incompletely [2]. Local resection is the treatment of choice for penile schwannomas and regular follow-up is suggested. Some patients complained of dyspareunia or disturbance during sexual intimacy. For the purpose of symptomatic amelioration, a simple total resection of the tumor is indicated. However, no definitive evidence supports the hypothesis that the erectile dysfunction may be attributable to penile schwannoma.

Based on a clinical and cytogenetic point of view, we present a rare case of plexiform schwannoma occurring on the glans. Although it has an identical embryologic origin, plexiform schwannoma of the glans has not yet been mentioned in the literature, as compared with the infrequent cases arising from the clitoris. Schwannomas exhibit a variety of chromosomal imbalances, among which a tumor suppressor gene may play a pivotal role in the genesis of these kinds of tumors. Whether varying types of schwannoma represent biologically distinct entities or a different pathway for tumorigenesis remains to be determined.

References