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Research Article

Myointimoma of Penis: Review and Update

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Abstract

Myointimoma, which is also known as angiocentric myofibroblastic tumour, is a rare benign soft tissue tumour which is derived from intimal cells of the vascular spaces of the corpora cavernosa of the penis, histologically characterized by multinodular/plexiform myofibroblastic proliferation within the vascular spaces of cavernous bodies. Myointimoma can afflict children and adult males and it does manifest as a lump or mass on the penis which have been noticed recently over a few weeks and which recently has been increasing in size within the glans penis. Generally, patients who are afflicted by myointimoma tend to be asymptomatic apart from noticing a lump. Diagnosis of myointimoma can be established via pathology examination and immunohistochemistry staining studies and molecular and cytogenetics studies of biopsy specimens or excised specimens of the tumour.

Total excision alone is sufficient for the management of myointimoma of the glans penis because the tumour portends a benign indolent cause. Less than 30 cases of myointimoma of the penis have so far been reported in the global literature and for this reason, majority of clinicians in the world have never encountered a case of myointimoma of the penis before and they would tend not to be familiar with the manifestation, diagnostic features, treatment, and outcome of the tumour pursuant to its treatment. The important thing to appreciate is the fact that myointimoma of the penis which simulates more common lesions of the penis that requires careful pathology examination and a high-index of suspicion by the clinician and the pathologist to distinguish myointimoma of the penis from its simulants including: plexiform fibrohistiocytic tumour of penis, epithelioid haemangioendothelioma of penis, myofibroma of the penis, intravascular fasciitis of penis, nerve sheath tumour of penis, leiomyoma of penis as well as various other types of lesions afflicting the penis.

Keywords: Myointimoma of penis; angiocentric myofibroblastic tumour of penis; penile mass; glans penis; corpus spongiosum; biopsy; histopathology; immunohistochemistry; molecular and cytogenetics studies; biopsy; excision; indolent; simulators

Introduction

Myointimoma, which is also referred to as angiocentric myofibroblastic tumour, is a rare benign soft tissue tumour which is derived from intimal cells of the vascular spaces of the corpora cavernosa of the penis, histologically typified by multinodular/plexiform myofibroblastic proliferation within the vascular spaces of cavernous bodies [1]. The terminology myointimoma was first used by Fetsch et al. [2] in 2000 and myointimoma is recognized as a distinctive histopathology entity in the World Health Organization Classification of the Tumours of the Urinary System and Male Genital Organs in 2016 [3]. To the knowledge of the author, so far, less than 30 cases of the myointimoma of the penis had been described in the literature, of which only 10 in children and adolescents were reported.

Except for two small series [4] of cases that are based upon a retrospective re-evaluation of few decades stored slides of tumours, these are always isolated case reports. Considering the rarity of this tumour, the main importance is to differentiate myointimoma from other tumours of variable biological behaviour. Considering the rarity of myointimoma, it would be envisaged that majority of clinicians globally, would not have encountered a case of

myointimoma before, and they would also not be familiar with the diagnostic features, treatment and outcome following treatment of the tumour. The ensuing article on myointimoma of the penis is divided into two parts: (A) Overview which has discussed general aspects of myointimoma of the penis and (B) Miscellaneous narrations and discussions from some case reports, case series and studies related to myointimoma of the penis.

Aim

To review and update the literature on myointimoma of the penis.

Methods

Internet databases were searched including google; google scholar; yahoo; and PUBMED. The search words that were used included: Myointimoma of the penis; myointimoma of glans penis; and myointimoma of corpus cavernous. Twenty (20) references were identified which were used to write the article which has been divided into two parts: (A) Overview which has discussed general aspects of myointimoma of penis and (B) Miscellaneous narrations and discussions from some case reports, case series and studies related to myointimoma of the penis.

Result

Overview

Definition / General

a) It has been iterated that myointimoma is a medical terminology that refers to a benign myointimal proliferation with predilection to the corpus spongiosum of the glans penis.

Essential Features

a) Myointimoma is a benign mesenchymal tumour which tends to be found within the corpus spongiosum of the glans penis.

b) Myointimoma is an intravascular proliferation of the vascular intimal cells.

c) In myointimoma, the lesional cells are stated to stain with α smooth muscle actin but not desmin.

Terminology

The terminologies which had tended to be used for myointimoma include:

a) Penile myointimoma.

b) Myointimoma of the penis [5].

c) Myofibrolastic tumour of penis.

Epidemiology

The epidemiology of myointimoma had been summarized as follows:

a) It has been iterated that myointimoma is a rare tumour with about 28 cases reported in the literature [6].

b) It has been pointed out that myointimoma had previously been described as intravascular leiomyoma, leiomyomatosis, late-stage intravascular fasciitis, leiomyoma of the glans penis, solitary cutaneous myofibroma of the glans penis [7,8].

c) It has been documented that there is a wide age range of patients who had been reported to be afflicted by myointimoma and the reported ages had ranged from 2 years to 74 years.

Sites

a) It has been documented that myointimoma afflicts the Corpus spongiosum of the glans penis.

Pathophysiology

a) With regard to the pathophysiology of myointimoma, it has been iterated the pathophysiology entails intravascular proliferation from the intimal cells of the inner layer of the corpus spongiosum vasculature in the glans penis.

Aetiology

a) It has been iterated that myointimoma is a mesenchymal tumour which is not related to the status of circumcision, history of trauma or the presence of disease [9].

Clinical Features

The clinical features of myointimoma had been summarised as follows:

a) Myointimoma, manifests as a small, distinct, non-mobile, firm, painless nodule upon the glans penis [10].

b) It has been iterated that myointimomas are benign, rapidly growing lesions that remain stable for years, even following incomplete excision of the lesions.

Diagnosis

The diagnosis of myointimoma, has been summarised as below:

a) Laboratory tests, including urine analysis and ultrasound scan evaluation of the abdomen and the scrotum to be undertaken to exclude other potential causes [11,12].

b) Punch, incisional or excisional biopsy of the lesion on the glans penis had tended to be undertaken for histopathology examination which confirms the diagnosis.

Prognostic Factors

a) With regard to prognosis, it has been iterated that myointimoma portends a benign outcome with no recurrence, and that spontaneous regression may occur.

Treatment

a) It has been stated that the treatment of myointimoma entails simple excision of the penile lesion.

Gross Description

a) It has been iterated that macroscopy examination of myointimoma does demonstrate a firm white mass that measures between 0.4 cm and 1.9 cm [2,4,10].

Frozen Section Description

a) It has been pointed out that frozen section examination may demonstrate that the lesion may be present at the margin of surgical excision.

Microscopic (Histologic) Description

The microscopy examination features of specimens of myointimoma had been summarized as follows:

a) Microscopy examination of specimens of myointimoma demonstrates multi-nodular or plexiform pattern; composed of occlusive intravascular myointimal proliferation.

b) Microscopy examination of specimens of myointimoma demonstrates that the nodules contain spindle or stellate shaped cells embedded in abundant fibromyxoid matrix or sometimes chondroid matrix.

c) Microscopy examination of specimens of myointimoma demonstrates that the cells have long eosinophilic cytoplasmic processes, blunt ended nuclei, fine chromatin and juxtanuclear vacuoles.

d) Microscopy examination of specimens of myointimoma demonstrates foci of degenerative changes appear as ghost cells.

e) Microscopy examination of specimens of myointimoma demonstrates no cytologic atypia, nuclear pleomorphism, prominent nucleoli or mitoses.

f) Microscopy examination of specimens of myointimoma demonstrates residual smooth muscle bundles surrounding the tumour.

g) Microscopy examination of specimens of myointimoma demonstrates that the overlying skin may show slight hyperkeratosis [13].

h) Microscopy examination of specimens of myointimoma demonstrates no necrosis or significant inflammation.

Immunohistochemistry Staining Studies

Positive Stains

Immunohistochemistry staining studies of specimens of myointimoma demonstrates that the cells of the myointimoma exhibit positive staining for the following tumour markers:

a) Muscle specific actin.

b) Alpha smooth muscle actin.

c) Calponin.

Negative Stains

Immunohistochemistry staining studies of specimens of myointimoma demonstrates that the cells of the myointimoma exhibit negative staining for the following tumour markers:

a) Desmin (shows the residual native smooth muscle of the vessel walls).

b) CD34 / CD31 / factor VIII related antigen (highlight the penetrating capillaries and the residual endothelial cells).

c) S100 protein.

d) AE1 / AE3.

e) Verhoeff-van Gieson elastic histochemical stain (demonstrates the meshwork of the elastic fibres surrounding the nodules).

Differential Diagnoses

The differential diagnoses of myointimoma of penis has been summarised to include the following:

Plexiform Fibrohistiocytic Tumour

a) Not intravascular.

b) Common in extremities, trunk, head and neck.

c) Dimorphic population of cells.

d) Fascicles of spindle cells with no desmin reactive collarettes of residual native smooth muscles, which are seen in myointimoma.

e) Nodules of histiocytes, including osteoclast-like giant cells (CD68+ and CD163+).

Epithelioid Haemangioendothelioma

a) Neoplastic cells are more epithelioid with intraluminal vacuoles instead of intracytoplasmic juxtanuclear vacuoles.

b) CD34+, CD31+ and SMA-.

Myofibroma

a) Biphasic pattern of myoid nodules similar to myointimoma; however, there are hemangiopericytoma-like areas.

b) Can involve vessels but is not limited to intravascular lumina.

Intravascular Fasciitis

a) May overlap.

b) Intralesional inflammatory cells, mucoid pools.

Nerve Sheath Tumour

a) Diffuse nuclear S100.

Leiomyoma

a) Not intravascular.

b) Fascicular architecture not plexiform or multinodular pattern.

c) Fibromyxoid stroma is not a typical feature.

d) SMA+ and desmin+.

Miscellaneous Narrations from some Case Reports, Case Series, and Studies related to Myointimoma of Penis

Fetsch et al. in their study, detailed out the clinicopathological and immunohistochemistry features associated with 10 cases of a

distinctive myointimal proliferation which had involved the corpus spongiosum of the glans penis. The ages of the patients had ranged from 2 years to 61 years and their mean age was 29 years. The patients were presented with a mass that varied in size from 0.5 cm to 1.9 cm in greatest dimension. The process was said to be present from 4 days to more than 6 months before surgical intervention. In each case, microscopy examination demonstrated almost identical histology. There was a prominent, often occlusive, fibro-intimal proliferation with plexiform architecture which had involved the vasculature of the corpus spongiosum. The proliferation was noted to have consisted of stellate-shaped and spindled cells embedded in abundant fibromyxoid matrix. Occasional lesional cells had well-developed myoid characteristics with moderately abundant eosinophilic cytoplasm, blunt-ended nuclei, and juxtanuclear vacuoles. Foci which contained degenerative changes, including "ghost cell" morphology, were also found upon pathology examination.

The myointimal process was noted to be extensively immunoreactive for alpha-smooth muscle actin, muscle-specific actin (HHF-35), and calponin, but it was minimally reactive for the D33 and D-ER-11 desmin clones. In contrast, native vascular smooth muscle surrounding the proliferation exhibited strongly positive immunoreactive staining for all five markers. The myointimal cells were nonreactive for CD34, S-100 protein, and keratin. Factor VIIIrAg, CD31, and CD34 highlighted intact endothelial cells lining sub-occluded vessels, scattered capillaries that penetrated the proliferation, and the normal uninvolved vasculature. The examined specimens were punch, incisional, or excisional biopsies, and in each instance, the process microscopically had extended to the tissue margin. Follow-up data were available for 8 cases in which the median follow-up interval, was 5 years and 8 months and one incompletely excised lesion with 6 months follow-up was noted to be stable but persistent, one lesion with 10 years follow-up had regressed spontaneously after a punch biopsy, and the remaining six lesions had not recurred.

McKenney et al. stated the following:

a) Penile myointimoma is a rare benign myointimal proliferation which occurs exclusively within the corpus spongiosum of the glans penis and is most commonly described in adult patients.

b) Up to the time of the report of their article. there was only one reported series of 10 penile myointimomas plus one case report, representing a total of 8 adults and 3 children/adolescents.

McKenney et al. reported 5 penile myointimomas which had occurred in 5 patients, who were less than 18 years of age and their ages had ranged between 4 years and 15 years. All manifested with a mass lesion upon the glans penis which had ranged in size from 0.4 cm to 1.8 cm. All 5 lesions had the classical morphology appearance: myointimal proliferation of the preexisting vascular spaces of the corpus spongiosum, creating a multinodular/plexiform architecture. Immunohistochemically, all stained cases showed strong cytoplasmic immunoreactivity for smooth muscle actin in the lesional cells and a collarette of native smooth muscle highlighted by desmin. None of the lesions had appeared completely excised, but all 5 patients were clinically free of disease at their last clinical follow-up and their follow-ups had ranged between 2 months and 45 months. McKenney et al. summated their report as follows:

a) They had reported only the second series of this distinctive, relatively rare myointimal proliferation within the corpus spongiosum of the glans penis, and they had expanded the number of published cases occurring in the paediatric/adolescent population, as well as they had confirmed the benign clinical course after a marginal or incomplete excision.

Casa et al. stated the following:

a) Penile myointimoma is a rare, benign tumour which occurs within the corpus spongiosum vasculature of the glans penis.

b) Up to the time of the report of their article, there had been twenty-three reported tumours in the literature.

Casa et al. presented four additional tumours of this unique myointimal proliferation. The ages of the patients had ranged from 20 years to 68 years and the patients had manifested with a firm mass on the glans penis. All four tumours had displayed distinctive morphological features consisting of a myointimal proliferation with plexiform architecture of bland myofibroblastic cells in a myxoid background in the corpus spongiosum vasculature. Characteristic cytoplasmic immunoreactivity of lesional cells with smooth muscle actin in addition to a desmin positive collarette of native vessel smooth muscle was seen in all four tumours. No disease recurrence was reported in any of the patients at their last clinical follow-up which had ranged between 9 months and 15 years, pursuant to their biopsy or excision.

Casa et al. made the ensuing conclusions:

a) Myointimoma is part of a rare group of mesenchymal tumours which had been recently classified by its distinctive location, morphology, and immunohistochemical reactivity.

b) For any nodular, spindle cell lesion of the corpus spongiosum, myointimoma should be included in the differential diagnosis of myointimoma given its unique characteristics and favourable clinical outcome.

Val-Bernal et al. described a cutaneous myofibroma with a monophasic pattern. Val-Bernal et al. reported a patient who was a 12-year-old white boy with an asymptomatic yellow nodule upon his glans penis for about 2 and half months. The reported nodular pattern of the growth was distinctive which comprised of nests of plump spindle cells with a tendency toward interstitial hyalinization within the dermis. Confirmation of the myofibroblastic nature of the proliferation was established by histochemical and immunohistochemical studies. The tumour cells exhibited immunoreactivities for vimentin, alpha-smooth-muscle actin, and collagen type IV but did not stain for desmin.

Val-Bernal et al. [14] stated the following:

a) To the best of their knowledge, this case was the first case of cutaneous myofibroma to occur on the glans penis.

b) This tumour should be included in the differential diagnosis of mesenchymal neoplasms of the penis.

Vardar et al. stated that myointimoma is a recently described benign tumour, which is regarded as a rare type of mesenchymal tumour of the penis. Vardar et al. reported a patient who was a 50-year-old man and who had a nodule located within his glans penis. He had a 2-month history of a mass. An excisional biopsy was undertaken. The histopathology examination findings demonstrated a multinodular tumour which was typified by spindle-shaped cells located in the intravascular area.

Vardar et al. stated the ensuing:

a) Their reported case, in addition to 11 cases that had been reported in the literature, had demonstrated that the myointimoma is frequently misdiagnosed on clinical and pathological grounds because of its rarity.

Tanriverdi, et al. stated the following:

a) Soft tissue tumours of the glans penis had been stated to be uncommon lesions.

b) One of these tumours is myointimoma which is quite rare.

c) It is a benign soft tissue tumour which is derived from the intimal cells of vessels within the corpus spongiosum of the glans penis.

d) The terminology "myointimoma" was utilised firstly by Fetsch for distinctive myointimal proliferation involving the corpus spongiosum.

e) There are few reports about this pathology.

f) They had reported an 11-year-old boy who had a mass, which was described as myointimoma, within his glans penis.

Tanriverdi, et al. reported an 11-year-old boy, who was referred to Manisa Celal Bayar University Hospital with a nodule within the left side of his glans penis. The nodule was detected two weeks earlier, and the patient had been asymptomatic except for the presence of the mass. The mass was located within the left side of his glans penis, and it measured 10 mm in size (Figure 1). The mass was also immobile and painless. The patient was circumcised when he was 6 years old. He did not have any history of trauma or infection. The results of his routine laboratory tests were within normal limits. He had ultrasound scan evaluation of his abdomen, urinary tract, and scrotal contents which was normal. The tumour was excised totally, and his glans penis was sutured primarily. It had an irregular border (Figure 2). Histopathology examination demonstrated that the mass had consisted of thick, smooth muscle fibres (Figure 3). There were focal fibrotic/fibromyxoid areas and spindle cell proliferation among them (Figure 4). The tumours cells were strongly stained with actin and weakly stained with desmin by immunohistochemistry (Figure 5). All these pathological findings were adjudged to be commensurate with the diagnosis of myointimoma. In the first year of follow-up, the patient did not have any problem regarding his penis or other urinary tract organs.

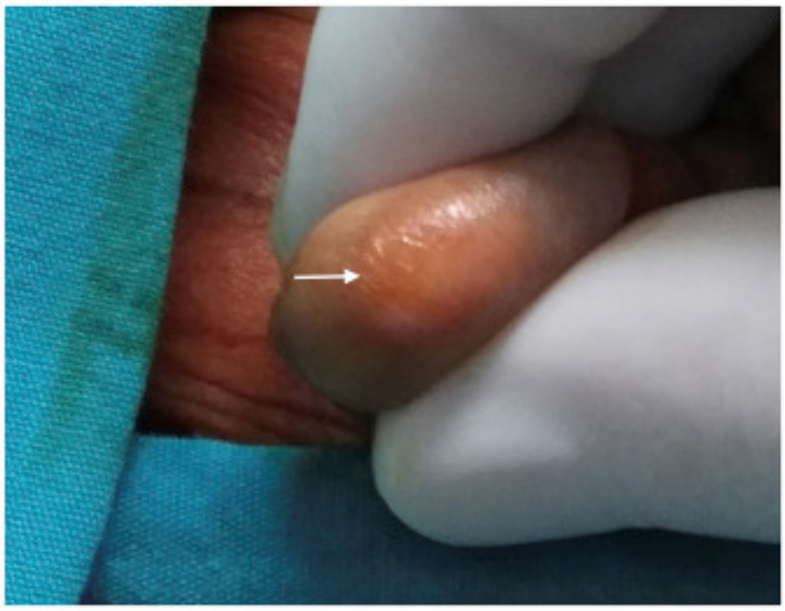

Figure 1: The nodule in the left side of glans penis (white arrow) [12].

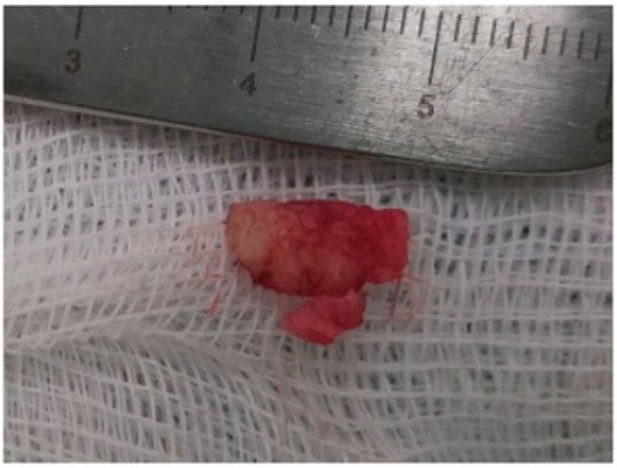

Figure 2: The nodule excised totally [12].

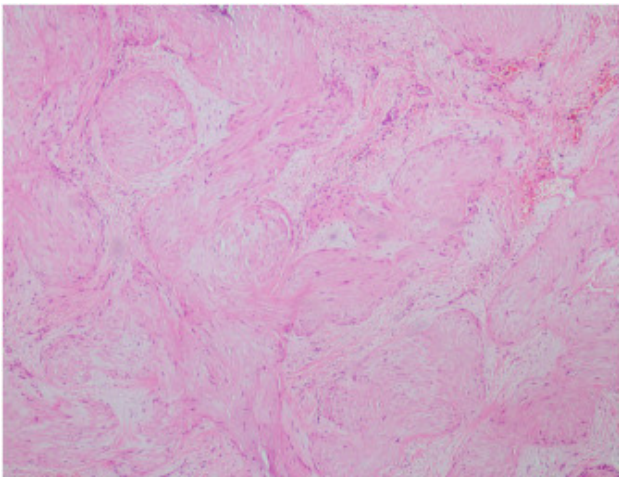

Figure 3: Thick, smooth muscle bundles (HE, x100) [2].

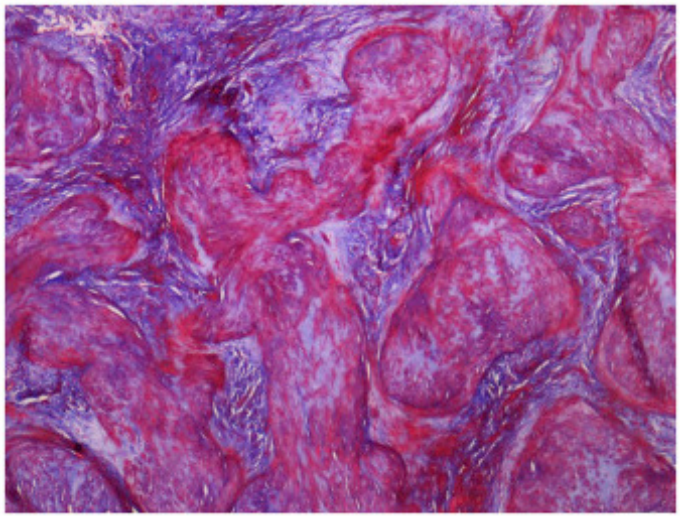

Figure 4: Focal fibrotic/fibromyxoid areas among smooth muscle fibres (Masson-trichrome, x100) [12].

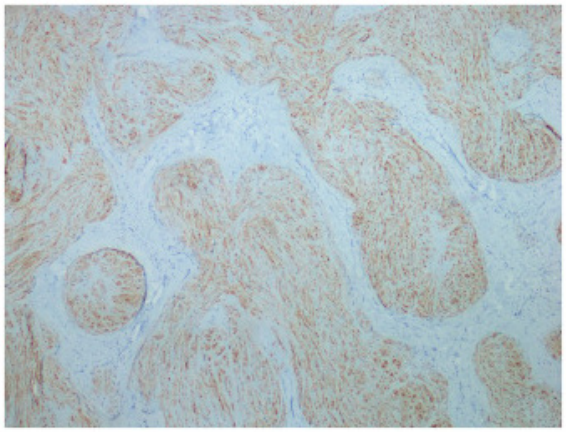

Figure 5: Strongly stained with actin (Actin, x100) [12].

Tanriverdi, et al. made the ensuing educative summating discussions:

a) Primary benign penile soft tissue tumours are unusual tumours.

b) Leioyoma, haemangioma, haemangioendothelioma, myofibroma, neurofibroma, schwannoma, may be seen within the penis mostly.

c) Myointimoma is a rare tumour which had recently been described as a benign soft tissue tumour of the penis.

d) It was described by Fetsch and there had been a few reports of myointimoma of the penis.

e) The age distribution of patients is wide.

f) Fetsch reported 10 cases and the ages of the patient had ranged between 2 years and 61 years.

g) McKenney reported 5 cases aged between 4 years and 15 years.

h) The tumour usually becomes markedly enlarged within a short time of about 1 month to 2 mounts.

i) Also, patients tend to be asymptomatic except for presence of the mass.

j) The lesions were sized from 0.4 cm to 1.9 cm, and they were located within the glans penis and corona in different studies.

k) Their patient was 11 years old. The nodule was detected two weeks preceding his presentation and he was asymptomatic except for presence of the mass. The nodule was approximately 10 mm in size.

l) Total excision of the nodule is enough treatment.

m) Local recurrence or metastasis had not been reported.

n) On the other hand, a spontaneous regression of the tumour after biopsy had been reported.

o) They had excised the mass totally and there was no recurrence within the glans for a year.

p) Typical histopathology examination of myointimoma findings include multinodular or plexiform intravascular myointimal proliferation of spindle cells within the glans penis.

q) Immunohistochemistry staining studies do demonstrate that myointimal cells exhibited positive immunoreactive staining for smooth muscle actin, muscle specific -actin, calponin and they exhibited negative staining for S-100 protein and desmin.

r) Myointimal proliferation is demonstrated with Verhoeff-van Gieson's stain as elastic fibres encompassing nodules.

s) In their patient there were thick smooth muscle fibres on pathological examination concordant with myointimoma.

t) It has been recommended that Myointimoma must be differentiated histologically from myofibroma, nodular fascitis, leiomyoma, plexiform fibrohistiocytic tumour, and epithelioid haemangioendothelioma.

u) Myoid predominant myofibromas could be nodular, but they do not demonstrate the exclusive intravascular growth like myointimoma.

v) Also, they do not demonstrate distinctive smooth muscle collarettes.

w) Fasciitis typically contains intralesional inflammatory, mucoid pools and less compact stroma.

x) Myofibroblastic proliferation within myointimoma is homogeneous and there are no composite lesions with hyalinized and loose stroll patterns of fasciitis.

y) Leiomyomas do not grow in a barbarizing intravascular pattern. They also have more well-developed fascicular structure and do not typically have a prominent myxoid stroma.

z) In children, plexiform fibrohistiocytic tumour is important for differential diagnosis because it might recur and it is associated with a small risk development of metastases.

aa) Even though both of the two lesions have a multinodular-plexiform appearance, plexiform fibrohistiocytic tumour is not an intravascular lesion.

bb) Dence myxoid stroma, cytoplasmic eosinophilia and intravascular location of myointimoma may simulate epithelioid haemangioendothelioma.

cc) Immunohistochemistry staining studies could be utilised for differential diagnosis in cases.

dd) A vascular lesion could be verified with CD31 - CD34 reactivity and absence of staining with smooth muscle-actin.

Tanriverdi, et al. concluded that:

a) They had presented a rare benign tumour called myointimoma within the glans penis.

b) It must be considered in the differential diagnosis, and it is well-known that total excision is sufficient treatment.

Cito et al. stated the following:

a) Myointimoma is an uncommon, benign soft-tissue tumour which is derived from the intimal cells of blood vessels.

b) Considering that little is known about this rare tumour entity, their aim was to describe an additional case and to perform the first literature review on this topic.

Cito et al. reported a 49-year-old Caucasian man, who had presented with a 12-month history of a palpable, firm, solitary lesion which had involved his glans penis. Upon clinical examination, there was a 1 cm palpable, endophytic well-circumscribed nodule located on the left side of his glans penis, close to the coronal sulcus, with disease-free external urethral orifice. The patient underwent complete excisional biopsy. A skin rhombus that measured 1.1 cm × 0.8 cm × 0.3 cm was removed and the biopsy sample, was fixed in 10% formaldehyde, and sent to the Pathology department. At his 18-month follow-up visit, the patient was clinically disease free. Histopathology examination of the specimen demonstrated a multinodular intravascular proliferation of the corpus spongiosum. This myointimal proliferation comprised of bland predominantly spindle cells in an abundant fibromyxoid stroma. Immunohistochemistry staining for smooth muscle actin (1A4), cytokeratins (AE1/AE3, CAM5.2), and CD34 were undertaken using the avidin-biotin complex (ABC) immunoperoxidase method. The lesional cells exhibited positive staining for smooth muscle actin and negative staining for cytokeratins and CD34. Cito et al. [15] additionally stated the following:

a) Myointimoma is confirmed to be a penile benign lesion that may be adequately treated with excisional biopsy.

b) Even after incomplete or marginal removal, the penile lesion had been demonstrated to remain stable overtime or regress.

c) Differential diagnosis is essential to exclude similar histologic entities that could be more aggressive or have possible systemic implications.

Drlik et al. reported a 15-year-old Caucasian boy, who had manifested with a 6-months history of a slowly growing, palpable firm nodule within his glans penis. Clinically he was completely asymptomatic and had passed urine freely. He did not report any history of trauma, systemic connective tissue diseases or other autoimmune disorders. Upon his clinical examination, there was a palpable, well circumscribed, firm, whitish painless mass, about 1 cm in diameter within his glans penis (Figure 6). His overlying skin was of a normal structure without signs of inflammation. No palpable inguinal lymphadenopathy was identified. His stage of puberty was Tanner III. Considering that there were no guidelines concerning penile tumours in this age, the authors adhered to the EAU guidelines for penile cancer in adults and undertook penile Doppler Ultrasound scan and MRI (Magnetic Resonance Imaging) scan.

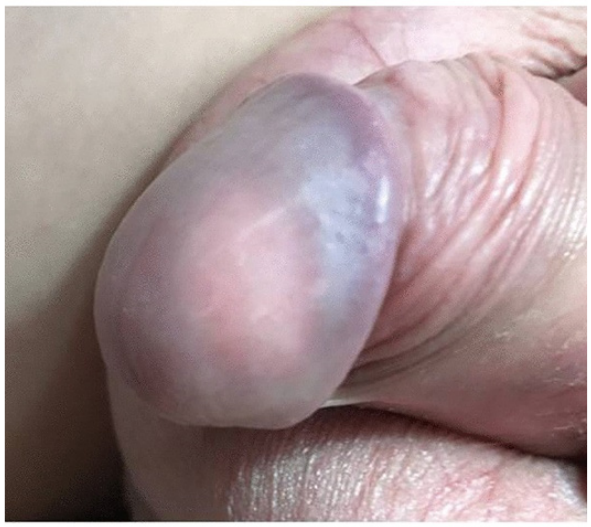

Figure 6: Whitish nodule visible under normal overlying skin [1].

The ultrasound scan demonstrated a hypoechogenic, hypo-perfused poorly defined area inside his glans (Figure 7). The MRI scan did not confirm any other pathological mass within the glans penis and corpora cavernosa (Figure 8). An excisional biopsy under general anaesthesia with intra-operative pathological evaluation was decided. The formation was not clearly demarcated from the encompassing glans tissues and had reached close to the urethra, without interfering with its wall. The procedure was undertaken at optical magnification, utilising magnifying glasses with particular attention to prevent injury to the neighbouring urethra (Figure 9). As the intra-operative pathology evaluation demonstrated a benign nature of the tumour, the authors simply closed the wound and did not proceed with any more extensive surgery (Figure 10). The authors obtained a macroscopically pale tissue sample that measured 10 mm × 8 mm × 5 mm (Figure 11).

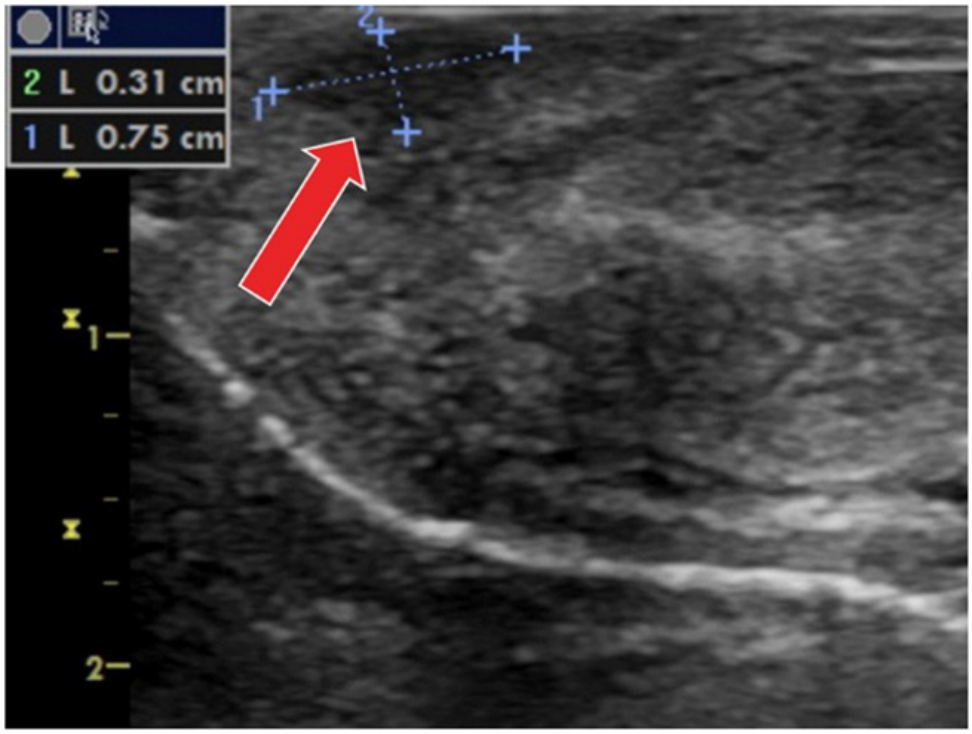

Figure 7: Ultrasound finding—a hypoechogenic, hypo-perfused non-well-defined area inside the glans (arrow) [1].

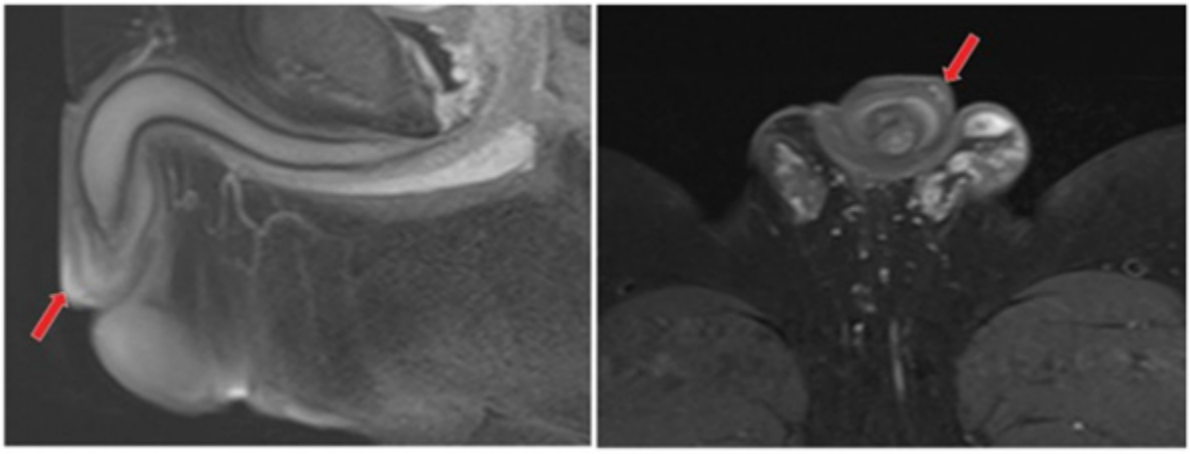

Figure 8: MRI finding—a single hyperintense mass inside glans (arrow), corpora cavernosa was normal, sagittal (A) and coronal (B) cut [1].

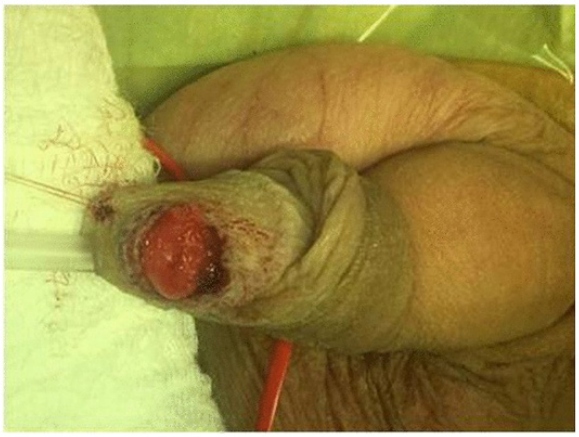

Figure 9: Careful excisional biopsy with special attention to protection of the urethra [1].

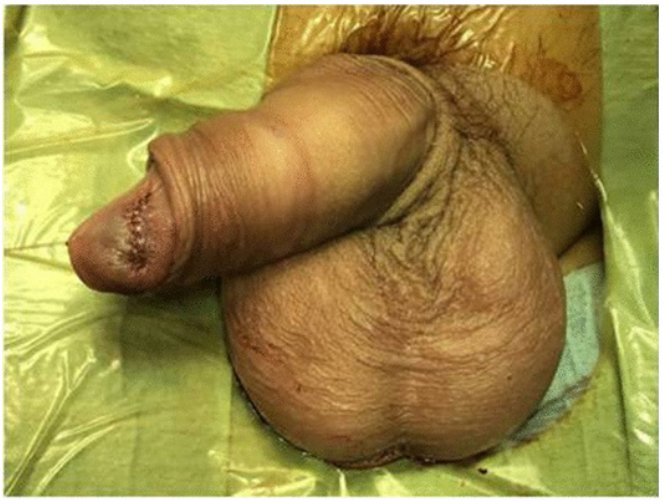

Figure 10: Simple wound closure [1].

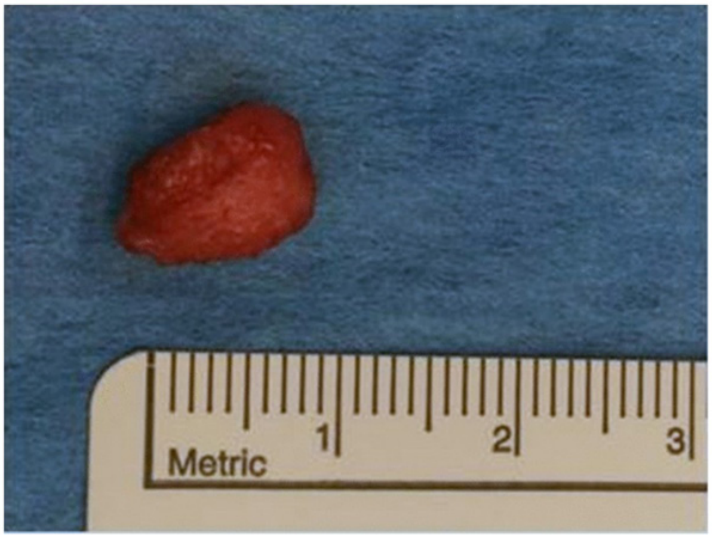

Figure 11: Macroscopic appearance of myointimoma [1].

examination of the specimen demonstrated changes diagnostic for myointimoma - nodular intravascular myofibroblastic proliferation which had involved multiple cavernous spaces (Figure 12). At low power magnification, a complex multinodular architecture was visualized. At higher magnification the myofibroblasts were found to be uniform, elongated spindle shaped cells with no significant hyperchromasia or pleomorphism, nor any mitotic figures or necrosis. Immunohistochemical staining for alpha-smooth muscle actin (αSMA) was positive intralesionaly (Figure 13), proliferative activity (Ki-67) was low (beneath 1%) (Figure 14). Immunohistochemistry staining for desmin was negative within myofibroblasts, while positive in the pre-existing vessel wall only (Figure 15). No reactivity was visualized for other performed immunohistochemical markers (S100 protein, CD34 and ERG). Due to the benign nature of the lesion, the authors did not undertake staging for distant metastases and simply undertook an outpatient follow-up. Three years after the excision, there was no local recurrence, no urethral stricture and a cosmetic appearance is good (Figure 16).

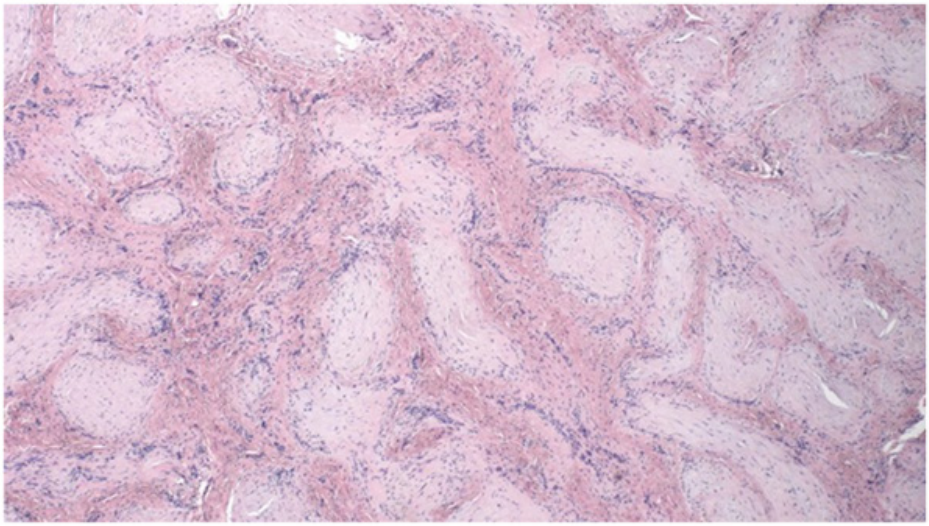

Figure 12: Nodular intravascular proliferation of spindle myofibroblastic cells presenting typical

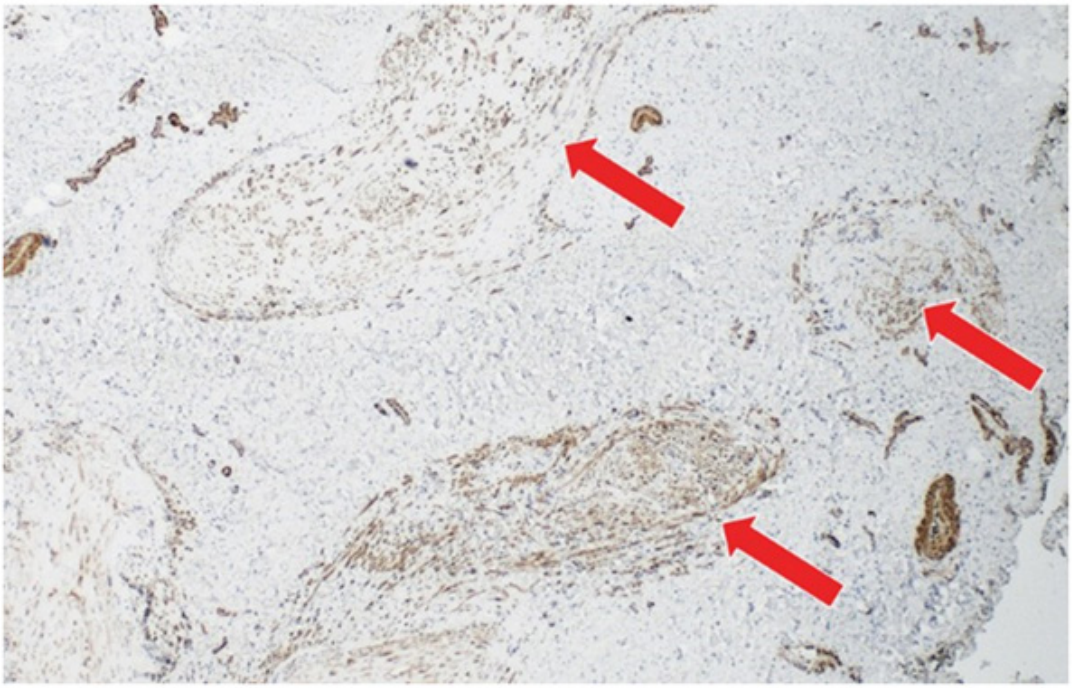

Figure 13: Immunohistochemical expression of alpha-smooth muscle action (SMA) shows diffuse positivity in intravascular myofibroblastic population (arrow) (200x) [1].

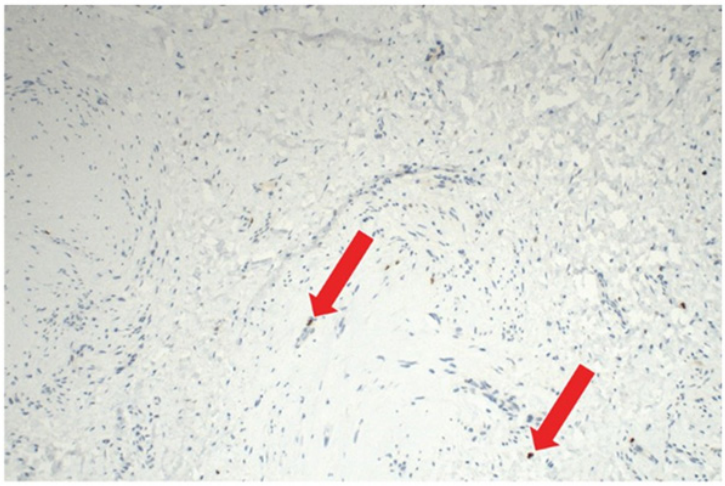

Figure 14: Immunohistochemical expression of Ki-67 (proliferative antigen MIB-1), labelling cells beyond G0 phase of the mitotic cycle, shows expression of sporadic cells (beneath 1%, arrow) (400x) [1].

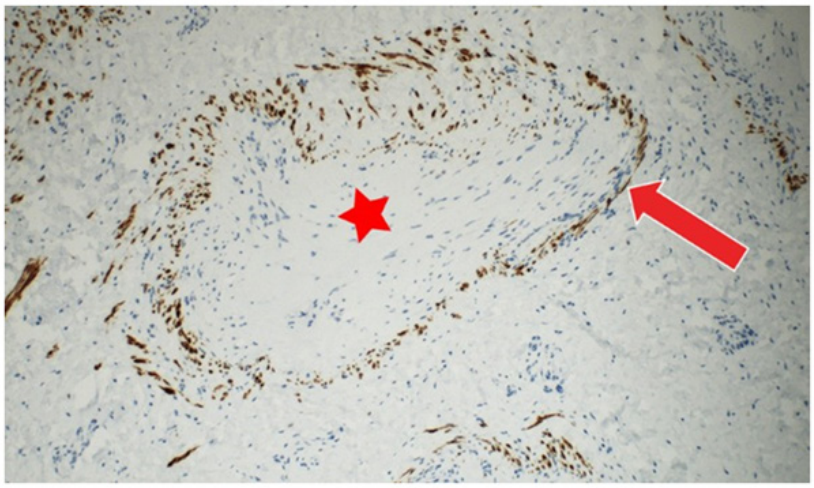

Figure 15: Immunohistochemical expression of desmin showing positivity in the smooth muscle cells of the pre-existing vessel walls (arrow), tumorous myofibroblastic cells are negative (star) (400x) [1].

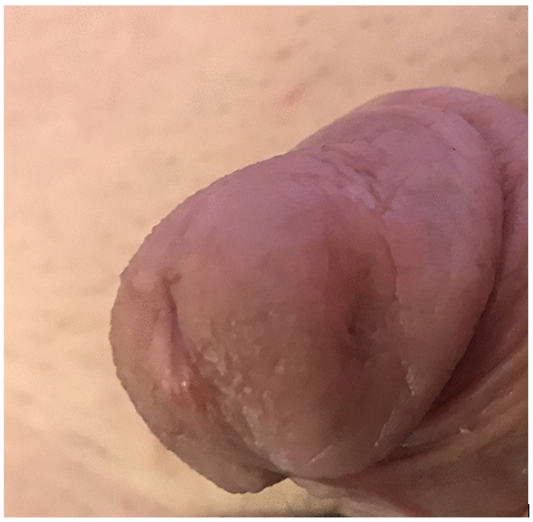

Figure 16: Favourable cosmetic outcome 3 years later [1].

adolescents.

b) With regard to their case, like in all previously reported cases in adolescents and adults [16], the myointimoma affects uniquely the glans penis.

c) Likewise, no reported case had been associated with pain, dysuria or signs of lower urinary tract obstruction.

d) In their reported case, the patient reported a relatively fast-growing mass.

e) The history of initial rapid growth is common in the reported literature; later nevertheless, the formation may remain stable over time.

f) Monsalves had reported a case of myointimoma which remained unchanged 10 months after an incomplete excision.

g) Fetsch reported the same experience with a 6-month stable residual mass in a patient pursuant to an incisional biopsy.

h) In their reported case, complete regression of myointimoma at 10-years follow-up was reported.

i) Local aggressive growth or distant metastases had never been reported.

j) At the time of the report of their case, there were no guidelines describing the extent of radiology imaging in adolescents with penile tumours.

k) The existing literature had not delt with the scope of radiology imaging; both existing series, of cases were based on a retrospective re-evaluation of stored hematoxylin and eosin-stained slides of penile tumours over the preceding few decades only.

l) Therefore, they had adhered to EAU guidelines for penile cancer in the adults and had undertaken penile Doppler Ultrasonography and MRI to exclude corporal invasion.

m) The examinations had confirmed the solid nature of the tumour and had excluded cystic lesion and multiple involvement of cavernosal tissue.

n) In a case of penile tumour in adolescents, the main concern was to exclude clinically aggressive conditions, thus an excisional biopsy was decided.

o) In view of the fact that the boy was confirmed to have benign findings on histopathology examination and had clinically normal findings on the inguinal nodes, they did not undertake staging (CT scan of thorax, abdomen, and pelvis).

p) The diagnosis of myointimoma and its differential diagnosis based upon morphology only might be confusing.

q) There are many types of mesenchymal tumours with plexiform or nodular structure.

r) Immunohistochemistry staining studies is a key to exact diagnosis. Myointimomas exhibit always positive expression for alpha-smooth muscle actin (αSMA). Desmin may be absent or show only focal reactivity.

s) There is no reactivity for S-100 protein, CD31, CD34, ERG, epithelial membrane antigen (EMA) or neuron specific enolase (NSE).

t) The plexiform growth pattern could be found in plexiform histiocytic tumour (PFHT) [17]. Unlike myointimoma, it does contain an admixture of two components: a differentiated spindle fibroblastic/myofibroblastic cells and a round histiocytic cell component containing multinucleated giant cells (osteoclast-like giant cells). Immunohistochemically, the histiocytes and multinucleated giant cell express CD68, whereas the spindle cells express αSMA. PFHT might recur and has a low risk of metastases (lymph node, lung). A plexiform or nodular growth pattern could be visualised in some nerve sheet tumours such as plexiform schwannoma [18] or neurofibroma. Immunohistochemical expression for S-100 protein is then helpful in the differential diagnosis.

u) The myointimoma structure may simulate myofibroma, which is a more common tumour in children. In contrast, it does not exhibit the exclusive intravascular growth; the growth rather tends to be concentric around the small vessels. The tumour is comprised of oval or spindle myoid cells [19].

v) Myopericytomas typified by a distinctive biphasic growth pattern, with central hypercellular zone composed of spindle tumour cells, hyalinization and myoid cell nodules visible towards the periphery of the tumour. In contrast to myofibroma, intravascular growth is more common in myopericytoma, but it does not indicate a malignant neoplastic process [20].

w) Epithelioid haemangioma and haemangioendothelioma could be differentiated from myointimoma by immunohistochemistry staining as the endothelial nature of the lesional cells could be confirmed by CD31, CD34 and ERG positivity.

x) Another structurally similar pathology is a late phase of intravascular fasciitis (intravascular nodular fasciitis). It has been stated that histologically, intralesional inflammatory cells between spindle myofibroblast cells, mucoid pools, a less compact stroma with more eosinophilic hyalinization, and obvious mitotic figures were observed.

y) Intravascular spindle cells lesion such as intravascular leiomyoma or leiomyomatosis could be easily differentiated by immunohistochemistry staining studies, with αSMA, desmin and h-caldesmon antibodies, which are typically strongly positive.

z) Last but not least, the possibility of sarcoma with angio-invasive spread should be excluded during the histopathology examination of the tumour.

aa) Both the clinician and pathologist need to be aware of this rare benign entity.

bb) The key to a correct diagnosis is based upon a careful histological examination of the specimen, including immunohistochemistry.

cc) Local excision is a safe and effective treatment modality.

Conclusion

a) Myofintimoma of the penis is a rare benign tumour of the penis.

b) Less than 30 cases of myointimoma of the penis has so far been reported in the global literature.

c) It is important to differentiate from more clinically aggressive neoplasms.

d) The key to a correct diagnosis of myointimoma of the penis is a careful histopathology examination, including immunohistochemistry staining studies of biopsy or excised specimen of the tumour.

e) Local excision of the tumour is a safe and effective treatment option that produces good outcome with no subsequent recurrence even in scenarios where there is tumour at the surgical resection margin.

f) Spontaneous regression of myointimoma of the penis has also been reported in a case of myointimoma of the penis which was diagnosed based upon pathology examination of a biopsy specimen of the tumour and in which the tumour was left alone without excision.

Conflict of Interest

None.

Acknowledgements

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