Aggressive Angiomyxoma of the Penis: The First Case Report in a 9-Month-Old Infant

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Aggressive angiomyxoma (AA) is a rare, benign, mesenchymal tumor of the pelvis and perineum. It usually occurs in females at reproductive age. However, rare cases have been reported in male children. We present the first case of AA in the penis of a 9-month-old Somalian boy. The infant presented with large, nodular penile mass. The diagnosis was confirmed following complete surgical removal and histopathologic examination. Over a 6-month follow-up period, no recurrence was noticed. Therefore, we recommend complete surgical removal of the mass with wide safety margin and long-term follow-up for cases of AA. UROLOGY 104: 187–190, 2017. © 2017 Elsevier Inc.

Angiomyxoma is a rare soft tissue tumor that has 3 types: aggressive, superficial, and angiomyofibroblastoma.[1] Aggressive angiomyxoma (AA) was described for the first time by Steeper and Rosai in 1983 as a rare mesenchymal tumor that mostly appears in the pelvis and perineum of adult females.[2] Few cases have also been reported in males. The male-to-female ratio is about 1:6.6.[3] AA usually has local infiltrative nature and high recurrence rate (36%-72%).[4] The term "aggressive" indicates, mostly, the high rate of local recurrence.[5] Surgical resection of the tumor with wide free margin is the treatment of choice. Long-term follow-up is recommended because of the high potential of recurrence.[6] It is very rare in male children; however, 4 cases have been described in the literature: 2 cases were found in the spermatic cord[7] and 2 cases in the scrotum.[8]

A case of AA in the penis of a 9-month-old infant is presented herein. To the best of our knowledge, this is the first reported case of AA in the penis and in this young age.

CASE REPORT

On May 1, 2016, a 9-month-old Somalian male infant presented to the Egyptian hospital in Mogadishu with large penis for age. According to history taken from his mother, the pregnancy was uneventful, with ordinary antenatal and postnatal history. She was not hypertensive or diabetic, and the infant was completely normal at birth. However, the infant's penis started to progressively increase in length and diameter when he reached 3 months old. This was not associated with other constitutional symptoms such as fever, refusal of feeding, and decreased activity. Furthermore, there were no urinary symptoms such as crying or straining during micturition, frequency, and hematuria. Additionally, there were no symptoms suggesting malignancy, such as failure to thrive and progressive loss of weight. The family history was irrelevant.

The initial physical examination showed that the baby had a very large penis, about 12 cm in length. It was nodular, firm in consistency, non-tender, with slight curvature to the right side (Fig. 1A). The testes and scrotum were normal. No enlarged regional lymph nodes and no other abnormalities were detected.

The pelviabdominal ultrasound was normal. Ascending urethrography revealed dilatation of a long distal part of the urethra (Fig. 1B).

The decision was to perform surgical exploration. After the patient was prepped and draped appropriately, a 8Fr Nelaton catheter was inserted into the external meatus and advanced into the bladder (Fig. 2A). Thereafter, a skin circumferential incision was performed for complete degloving of the penis (Fig. 2A). During dissection, it became evident that the distal part was a thick tissue glistening mass and a normal-sized penis was found proximally (Fig. 2B). The mass was excised and sent for histopathologic examination and the penis was repaired as shown in Figure 2C,D.

Gross examination of the mass showed that it was about 12 cm in length, grayish-white in color, gelatinous, glistening, and firm in consistency (Fig. 2D). Microscopic examination of the specimen revealed a non-encapsulated, hypo/hypercellular proliferation tissue of penile tunica

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Figure 1. **(A)** Large nodular penile mass (about 12 cm), curved to the right side. **(B)** Ascending urethrography (right anterior oblique view) shows dilated distal part and normal proximal part of the urethra. (Color version available online.)

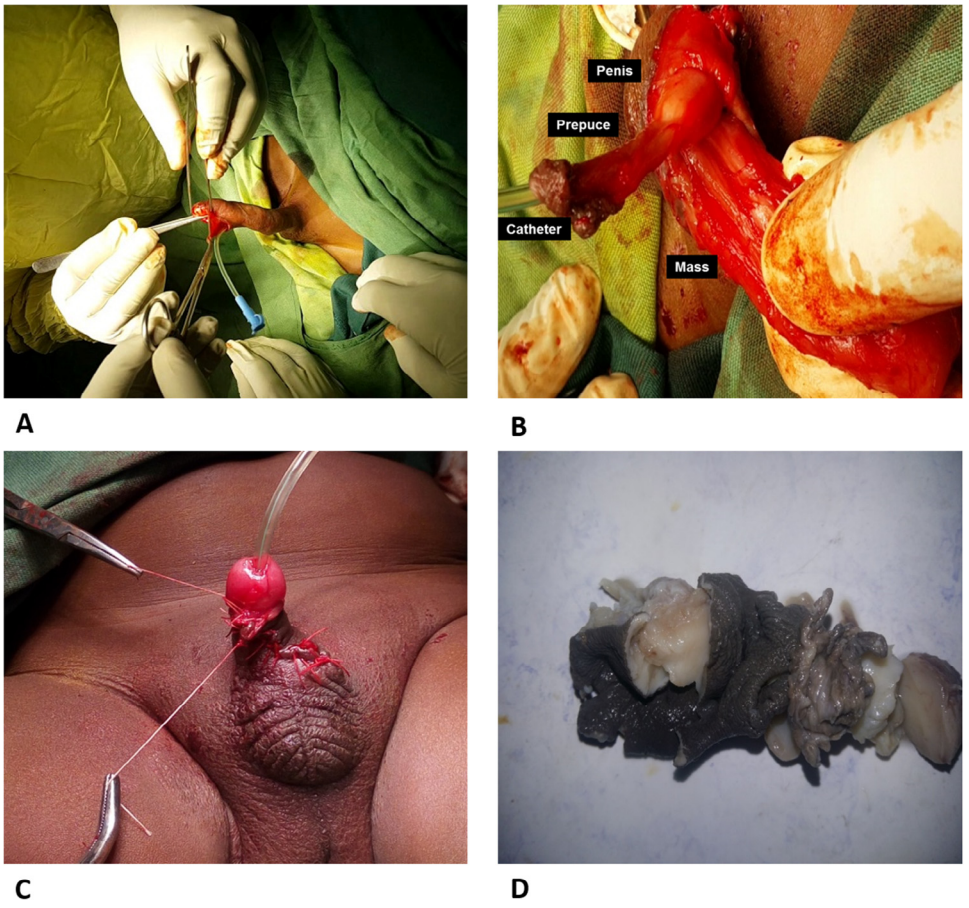

Figure 2. **(A)** Insertion of 8Fr Nelaton catheter and beginning of degloving of the penis. **(B)** Separation of the mass from the penis. **(C)** Final view for the penis following removal of the mass. **(D)** Gross picture for the removed mass. (Color version available online.)

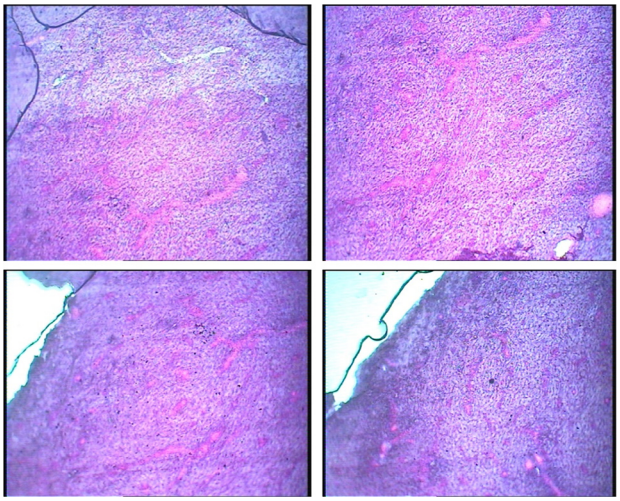

Figure 3. Microscopic picture shows proliferated spindle to stellate cells in loose myxoid stroma with variable caliber blood vessels. (Color version available online.)

albuginea cells, composed of widely dispersed, uniform population of spindle to stellate cells, with round to oval nuclei, in loose myxoid stroma. The blood vessels were of variable caliber and with partially thickened walls (Fig. 3).

After 6 months of follow-up, no recurrence was noted. The patient's family has been informed to keep the follow-up because of the high risk of recurrence.

DISCUSSION

AA, as a rare mesenchymal tumor, is uncommon in males and very rare in male children.[2] The male-to-female ratio is about 1:6.6.[3] Review of literature revealed only 4 cases of AA in male children: 2 cases in the spermatic cord[7] and 2 cases in the scrotum.[8]

To our knowledge, this is the first case of AA in the penis. Herein, we presented a case of a 9-month-old Somalian infant who presented with a 12-cm, nodular, non-tender penile mass. After surgical excision, histopathologic examination revealed non-capsulated mass composed of proliferated spindle to stellate cells with variable caliber, thick-walled blood vessels corresponding to AA.

Classically, the gross picture of AA is mostly a firm, nodular, glistening, and poorly circumscribed mass. Microscopically, it appears as a myxoid stroma with cellular proliferation of spindle to stellate cells, and blood vessels of variable size with partial thick walls.[7] This was congruent with the findings in our case.

AA should be differentiated from other types of angiomyxoma, such as superficial angiomyxoma and angiomyofibroblastoma. Superficial angiomyxoma is usually a subcutaneous multinodular mass, with small-sized, thin-walled blood vessels that lack the hypertrophic vessels and infiltrative nature of AA.[9] In addition, neutrophilic infiltration in superficial angiomyxoma is an important histologic clue for differential diagnosis with other myxoid lesions.[10,11] Recently, the first case of recurrent superficial angiomyxoma in an 18-year-old Chinese male patient was reported.[12] It occurred as 2 asymptomatic masses on the proximal and distal ends of the penis. The patient had a history of a solitary proximal penile nodular mass that recurred 6 months following surgical removal. After complete re-excision of the masses, gross and histopathologic examinations revealed that these masses were superficial angiomyxoma. No recurrence was reported after 80 months of follow-up. Unlike our cases, this case of recurrent superficial AA was accompanied with enlarged bilateral inguinal lymph nodes which disappeared after 8 months of complete excision of the both masses.[12]

Furthermore, angiomyofibroblastoma is composed of epithelioid cells distributed around the vessels,[8] and it differs from AA in its characteristic appearance and immunophenotype of the stromal cells.[9] AA should also be distinguished from other soft tissue tumors, such as myxoid neurofibroma, myxoma, spindle cell lipoma, and myxoid liposarcoma.[6]

To date, surgical excision, with wide free margins, is considered as the most effective treatment of AA. In addition, long-term postoperative follow-up is required because of the high risk of local recurrence.[6] The follow-up is best achieved by magnetic resonance imaging.[13] Recurrent cases need repeated surgical interventions.[4] However, hormonal treatment with tamoxifen, raloxifene, and GnRH analogs has been tried and resulted in decreasing the size of the tumor.[14]

In the current case, no evidence of recurrence was noted after a follow-up duration of 6 months. Because of the high risk of recurrence, the patient's family was advised to continue follow-up and they were advised that the infant might need another surgical intervention if recurrence occurred.

CONCLUSION

In this report, a rare case of AA in the penis of a 9-month-old male infant was described. Complete surgical removal of the mass with wide safety margin and long-term follow-up is recommended.

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