

# Aggressive Angiomyxoma of the Scrotum: A Case Report and Literature Review

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**Abstract** Aggressive Angiomyxoma is a rare mesenchymal benign myxoid tumor of the pelvis and perineum which occurs almost exclusively in adult females. It rarely occurs in males and involves scrotum, locally infiltrative but nonmetastasizing. The clinical presentation was one of progressing scrotal edema and enlargement with subsequent development of scrotal abscesses. The clinical differential diagnosis at presentation commonly includes inguinal hernia, testicular neoplasm, spermatic cord lipoma, hydrocele, spermatocele, and scrotal filariasis. We report a case of incidentally diagnosed Angiomyxoma of scrotum presenting as scrotal edema. **Case Report:** A 41 years old male presented with a painless scrotal swelling since 2 years prior to admission that gradually progressed in size. Clinical examination revealed diffuse scrotal swelling measuring about 15×12cm. with a thickened of scrotal skin. Ultrasound examination report was found of fluid collection with debris in the scrotum suspected as scrotal abscess, subcutaneous edema and cutaneous scrotal wall thickening. **Result:** The patient underwent wide excision and scrotoplasty, the pathology result of the excised scrotal edema was an Angiomyxoma of the scrotum up to tunica vaginalis. **Conclusion:** Aggressive Angiomyxoma in the scrotal region may present as a scrotal edema and enlargement, often mistaken for an inguinal hernia, testicular neoplasm, spermatic cord lipoma, hydrocele, spermatocele, and scrotal filariasis.

**Keywords:** Angiomyxoma, Scrotal Edema

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## 1. Introduction

Aggressive angiomyxoma was first described by Steeper and Rosai in 1983. [1] It is a rare mesenchymal benign myxoid soft tumor of the pelvis and perineum which occurs almost exclusively in adult females. [2]

Overall, its incidence is about 6-folds higher in females. [3-5] Reports of the Aggressive Angiomyxoma in males have nevertheless been rare. [6]

Aggressive angiomyxoma is usually arising from the soft tissues of the pelvic region, perineum, vulva and buttock. [4] It rarely occurs in males and involves scrotum, spermatic cord, inguinal region, and perineum, locally infiltrative but nonmetastasizing and has a high risk of local recurrence after excision. [7,8]

The adjective “aggressive” emphasizes the neoplastic character of the blood vessels, its locally infiltrative nature and the high risk of local recurrence after surgical excision. It is called Angiomyxoma because of its myxoid nature and prominent vascularity. [5,9]

The clinical presentation in males was one of progressing scrotal edema and enlargement with subsequent development of scrotal abscesses. The clinical differential diagnosis at presentation commonly includes inguinal hernia, testicular neoplasm, spermatic cord

lipoma, hydrocele, spermatocele, and scrotal filariasis. It can be confused with malignant neoplasms and need to be distinguished from malignant myxoid neoplasms. [2]

The tumors are characterized by local infiltration, a gelatinous cut surface, frequent recurrence, no metastasis, slow growth, large size, and finger-like projections, and are occasionally encapsulated. [2,3]

Microscopically, it shows hypocellular, myxoid tissue with numerous medium sized vessels. Tumour cells are spindle cells, fibroblasts and myofibroblasts, that lack significant nuclear atypia and mitotic activity. [5,7,10]

Here, we report a case of incidentally found aggressive angiomyxoma of scrotum presenting as scrotal edema. Then, we reviewed and compared this case with other literatures as a retrospective study, in order to evaluate this case and provide better pre operative work up, treatment, and follow up similar case in the future.

## 2. Case Report

A 41 years old male presented with a painless scrotal swelling since 2 years prior to admission that gradually progressed in size which has been regarded as lymphedema.

There were no history of trauma, fever, weight loss, or other genitourinary disease.

Clinical examination revealed diffuse scrotal swelling measuring about 15×12cm with a thickened of scrotal skin, non fluctuant, nontransluminant, and non-tender, conforming to a scrotal lymphedema was seen. Both testicles were unpalpable and no inguinal lymphadenopathy was found (Figure 1).



Figure 1. Clinical picture

Microfilarial test was negative and other laboratory findings were within normal limits.



Figure 3. Right and Left Testicle Ultrasound



Figure 2. Scrotal Ultrasound

Ultrasound examination report was found of fluid collection with debris in the scrotum suspected as scrotal abscess, subcutaneous edema and cutaneous scrotal wall thickening. No signs of hydrocele, both testis and epididymis were normal (Figure 2 and Figure 3).

The clinical impression was a lymphedema, scrotal exploration and excision of scrotum with wide margins was performed to relieve the patient of discomfort from the swelling, then the scrotum was reconstructed (Scrotoplasty) (Figure 4).



Figure 4. Post operative clinical picture

On scrotal exploration, the mass was found between the tunica vaginalis and the skin, which displaced the testis upward.



Figure 5. Excised Scrotal Mass

The mass had a smooth surface, lobulated and was encapsulated and its cut surface was gelatinous with slimy areas (myxoid areas) (Figure 5).

An excision of the scrotal swelling and biopsy of tunica vaginalis was done and sent for histopathological examination.

Histopathology feature of the scrotum up to tunica vaginalis revealed the mass is composed of spindle & stellate cells, with hypocellular and myxomatous fibrocollagen matrix and numerous of lymphovascular vessels, partially with thickening wall. There is no malignant cell found (Figure 6, Figure 7, and Figure 8).

The pathology result of the excised scrotal edema up to tunica vaginalis was an Angiomyxoma.

Based on these histopathology features, a diagnosis of Aggressive Angiomyxoma was made.

On follow up, there were no clinical signs of recurrence after 6 months.

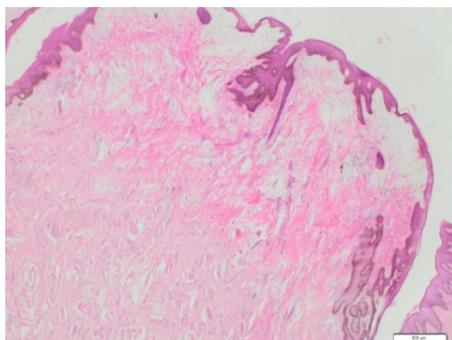


Figure 6. Histopathology result 5 X

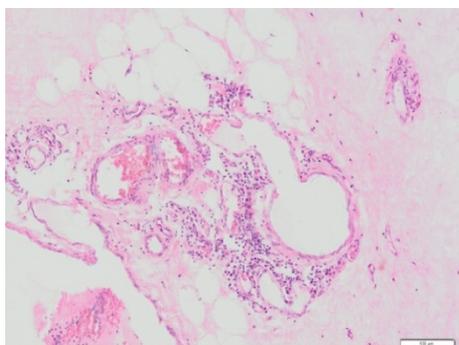


Figure 7. Histopathology result 10 X

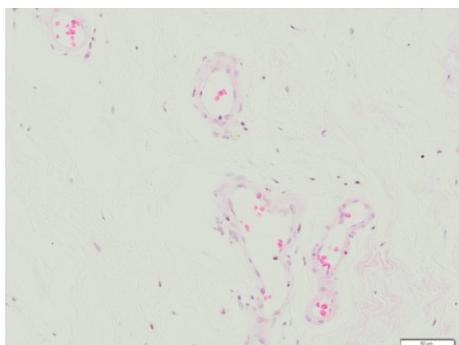


Figure 8. Histopathology result 50 X

### 3. Discussion

Since 1983, when aggressive angiomyxoma was first described by Steeper and Rosai, there were about 150 cases reported worldwide. [5]

It is a locally infiltrative mesenchymal tumor which is common in the perineal region in young females and is rare in males.

Aggressive Angiomyxoma occurs almost exclusively in the vulva, vagina, pelvic floor, and perineum of females. The tumor typically presents as an asymptomatic mass in females, and is sometimes confused with Bartholin's gland cysts or perineal hernias. [2,3]

Occurrence of aggressive angiomyxoma in males is extremely rare and in males, aggressive angiomyxoma is usually derived from the pelvipereineal interstitial tissue involving the scrotum (38%), spermatic cord (33%), perineal region (13%), and intrapelvic organs (8%). [3,7]

The adjective "aggressive" emphasizes the neoplastic character of the blood vessels, its locally infiltrative nature, and the high risk of local recurrence, not indicating a malignant potential of the lesion. And due to its locally infiltrative course and frequent recurrences it has been termed aggressive. [5,7]

Aggressive angiomyxoma in the scrotal region may present as a scrotal edema and enlargement, it tends to grow very slowly, and patients tend to demonstrate a prolonged period of disease progression. Preoperatively, diagnosis is difficult, often mistaken for an inguinal hernia, testicular neoplasm, spermatic cord lipoma, hydrocele, spermatocele, and lymphedema. [2,3,10]

Aggressive angiomyxoma should be distinguished from benign tumours with a low risk of recurrence such as intramuscular myxoma, neurofibroma, myxoid lipoma, spindle cell lipoma, angiomyofibroblastoma on one hand and from tumours with metastatic potential such as myxoid liposarcoma, myxoid malignant fibrous histiocytoma, embryonal rhabdomyosarcoma on the other. [10]

It is usually difficult to identify the tumor by imaging studies such as ultrasonography and there are reports showing some advantage to scrotal MRI as it better demonstrates the angiomatous and myxomatous nature of the tumor. However, most cases are currently visualized by CT scans which are more accessible for the patients. [3]

On CT scan, these tumors have a well-defined margin with attenuation less than that of the muscle. On MRI, these tumors show high signal intensity on T2-weighted images. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high water content of angiomyxoma. [7]

Ultrasound guided needle biopsy has been shown to give inconclusive diagnosis and the differential diagnosis includes angiomyofibroblastoma, myxoma and myxoid liposarcoma. Therefore, the final diagnosis usually awaits the final histological examination of the excisional specimen. [3]

Grossly, aggressive angiomyxoma is a grayish-colored, tumor with a smooth surface but is occasionally encapsulated. The tumor is soft, partly circumscribed and has a gelatinous appearance cut section. [2,5]

The characteristic histological findings are ill-defined margins, prominent vasculature of small to medium caliber with thin and thick walls, abundant myxoid matrix, spindle to stellate fibroblast-like cells, cytologically bland nuclei, few mitoses, and variable fibrosis. [2,5,7]

The thick walled vessels are therefore useful for differentiating between other soft tumors of the pelvis, including myxoid neurofibroma, myxoma, myxolipoma,

angiomyofibro blastoma, spindle cell lipoma, myxoid leiomyoma, myxoid leiomyosarcoma, myxoid malignant fibrous histiocytoma, myxoid liposarcoma, and Botryoid rhabdomyosarcoma. [2]

The tumor is usually locally infiltrative and has a high rate of local recurrence after surgical excision. The excision of these tumors is difficult as they have the same consistency as that of normal connective tissue and

therefore have a probability for local recurrence (36–72 %) that may be attributed to incomplete tumor resection. [7]

The mainstays of treatment are wide excision with tumor-free margins and close postoperative monitoring. Long-term follow-up remained necessary because of the risk of late recurrence. [2] MRI is the preferred method for detecting recurrences. [7] Metastases are exceedingly rare, and overall, the prognosis is good. [11]

**Table 1. Aggressive Angiomyxoma of the Scrotum: Literature review**

| Reference          | Age (year) | Pre operative work up                 | Working Diagnosis    | Treatment           | Size (cm)   | Recurrence    | Follow up periods |
|--------------------|------------|---------------------------------------|----------------------|---------------------|-------------|---------------|-------------------|
| Chuang et al. [2]  | 24         | AFP, $\beta$ hCG, Ultrasound, MRI     | Paratesticular tumor | Wide excision       | 7 x 5 x 5   | No            | 26 months         |
| Morag et al. [3]   | 64         | Ultrasound, CT                        | Hydrocele            | Scrotal exploration | 7 x 11 x 19 | No            | 3 years           |
| Chirara et al. [4] | 47         | AFP, $\beta$ hCG, Ultrasound, CT, MRI | Soft tissue tumor    | Extirpation         | 17 x 10 x 6 | No            | 14 months         |
| Padma et al. [5]   | 62         | Ultrasound                            | Scrotal edema        | Subtotal excision   | 15 x 12 x 8 | Not available | Not available     |
| Rao et al. [7]     | 62         | Microfilial test, Ultrasound          | Scrotal lymphedema   | Subtotal excision   | 15 x 12     | No            | 3 months          |
| Present case       | 41         | Microfilial test, Ultrasound          | Scrotal edema        | Wide excision       | 15 x 12     | No            | 6 months          |

Compared with other literatures, in this present case the aggressive angiomyxoma occurred in male and involving the scrotum up to tunica vaginalis. The clinical manifestation was similar with the other literatures as a progressing scrotal edema and enlargement, then we suspected this case as a scrotal filariasis. However, in this present case as it was a retrospective diagnosis, CT and MRI were not done preoperatively. On follow up, there were no signs of recurrence in clinical examination after 6 months following surgery.

Furthermore, the comparative pre operative work up, treatment, and follow up between present case with other literatures describes in Table 1.

## 4. Conclusion

Aggressive Angiomyxoma is a benign tumor and it must be considered in the differential diagnosis of scrotal edema and enlargement. It often mistaken for an inguinal hernia, testicular neoplasm, spermatic cord lipoma, hydrocele, spermatocele, and scrotal filariasis because of the lack of awareness of this entity. Detailed radiological work up may be helpful in detection, but histology is the gold standard for establishing the diagnosis. The choice of treatment are wide excision with tumor free margins and close post operative monitoring with long term follow up.

In this present case, the aggressive angiomyxoma of scrotum found incidentally after surgical treatment based on pathology result. The clinical examination presenting as a scrotal edema, and only using Ultrasound for imaging study. The treatment was wide excision, and we assume the tumor was completely excised since there were no signs of recurrence based on clinical examination after 6 months.

For further suspicious case alike, careful pre operative work up should be done, such as CT scan or MRI to detect angiomatous and myxoid components, and for planning of precise incision margin. Wide excision should be performed with tumor free margins as a treatment. On follow up, MRI should be carried out even if no signs of recurrence found in clinical examination.

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## References

- [1] Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum: Report of nine cases of a distinctive type of gynaecologic soft tissue neoplasm. *Am J Surg Pathol.* 1983; 7: 563-75.
- [2] Chuang FP, Wu ST, Lee SS, Chen HI, Chang SY, Yu DS, et al. Aggressive Angiomyxoma of the Scrotum: Case Report. *Archives of Andrology.* 2002; 48: 101-6.
- [3] Morag R, Fridman E, Mor Y. Aggressive Angiomyxoma of the Scrotum Mimicking Huge Hydrocele: Case Report and Literature Review. *Case Rep Med.* 2009; 2009: 157624.
- [4] Chihara Y, Fujimoto K, Takada S, Hirayama A, Cho M, Yoshida K, et al. Aggressive Angiomyxoma in the Scrotum expressing androgen and progesterone receptors. *Int J Urol.* 2003; 10: 672-5.
- [5] Padma M, Koteswari M, Rao NB, Sindu P, Lalithasree K. Aggressive Angiomyxoma of Scrotum, a Rare Entity: Case Report. *International Journal of Medical Science and Clinical Inventions.* 2015; 2: 633-5.
- [6] Iezzoni JC, Fechner RE, Wong LS, Rosai J. Aggressive angiomyxoma in the males: a report of four cases. *Am J Clin Pathol.* 1995; 104: 391-6.
- [7] Rao KSS, Rao KA, Praveen S. Aggressive Angiomyxoma of Scrotum Presenting as Scrotal Lymphedema in a Case of Postoperative Carcinoma Penis: Case Report. *Indian J Surg.* 2015; 10: 1007.
- [8] Kidric DM, MacLennan GT. Aggressive Angiomyxoma of the Male Genital Region. *The Journal of Urology.* 2008; 180: 1506.
- [9] Tsang WY, Chan JK, Lee KC, Fisher C, Fletcher CD. Aggressive Angiomyxoma: a report of four cases occurring in men. *Am J Surg Pathol.* 1992; 16: 1059-65.
- [10] Rao SN, Dais E, Prabhu S, Kumar TKS, Jose P. Aggressive Angiomyxoma of the Epididymis: Case Report. *Indianjurol.* 2001; 18: 96-7.
- [11] Sutton BJ, Laudadio J. Aggressive Angiomyxoma. *Arch Pathol Lab Med.* 2012; 136: 217-8.