Aggressive angiomyxoma of vulva - A case series

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Background

- Aggressive angiomyxoma (AAM) is a rare soft tissue neoplasm occurring mainly in young females.
- It was first described by Steeper and Rosai in 1983.
- Due to its rarity and presentation mainly as a labial cyst, it is often misdiagnosed.[1]
- It is a low-grade neoplasm with high local recurrence following incomplete excision because of its aggressive infiltrative nature.
- We report clinicopathological features of six cases of AAMs.

Material and Methods

- A retrospective observational study was conducted from 2012 to 2021.
- Patient details were obtained from the hospital database.
- All the slides and blocks were reviewed.

Results

- The mean age of presentation was 36 years.
- Two of the cases presented as recurrent mass following previous surgery.
- All the cases presented as vulval mass ranging in size from 5 to 26 cm.
- Grossly, the tumors are unencapsulated masses with tan pink to tan grey and gelatinous appearance.
- Microscopically, the tumor showed paucicellular areas composed of bland-looking oval to spindle cells set haphazardly in myxoedematous and collagenous stroma. (Fig 1 to 4)
- Many prominent vessels were also noted.
- Myoid differentiation was identified in four out of six cases.
- The two recurrent cases also shared similar morphology as the rest of the cases.
- At least one of the smooth muscle markers (calponin, desmin or smooth muscle actin) was positive in all the cases. (Fig 5)
- Estrogen receptors were positive in all cases (Fig 6) while progesterone receptor was negative in one case.
- The clinic-pathological features are shown in table 1.
- All the patients were disease-free and alive on follow-up.

	Age (years)	H/O recurrence	Imaging	Size (cm)	Morphology	IHC (positive markers)	Follow up
Case 1	23	Absent	CT	26x11x11	Classical+myoid differentiation	ER, PR, SMA	Alive
Case 2	36	Present	MRI	5x2.6x2.3	Classical+myoid differentiation	ER,PR, SMA	Alive
Case 3	37	Absent	MRI	11x10x9.8	Classical+myoid differentiation	ER, PR, SMA	Alive
Case 4	38	Present	MRI	6.1X4X3.5	Classical	ER, PR, Desmin	Alive
Case 5	30	Absent	MRI	7.5X4.3X4	Classical	ER, PR, SMA	Alive
Case 6	53	Absent	CT	5.3x4.6x4.3	Classical+myoid differentiation	ER,Calponin	Alive

Table (1) showing clinicopathological features of the cases

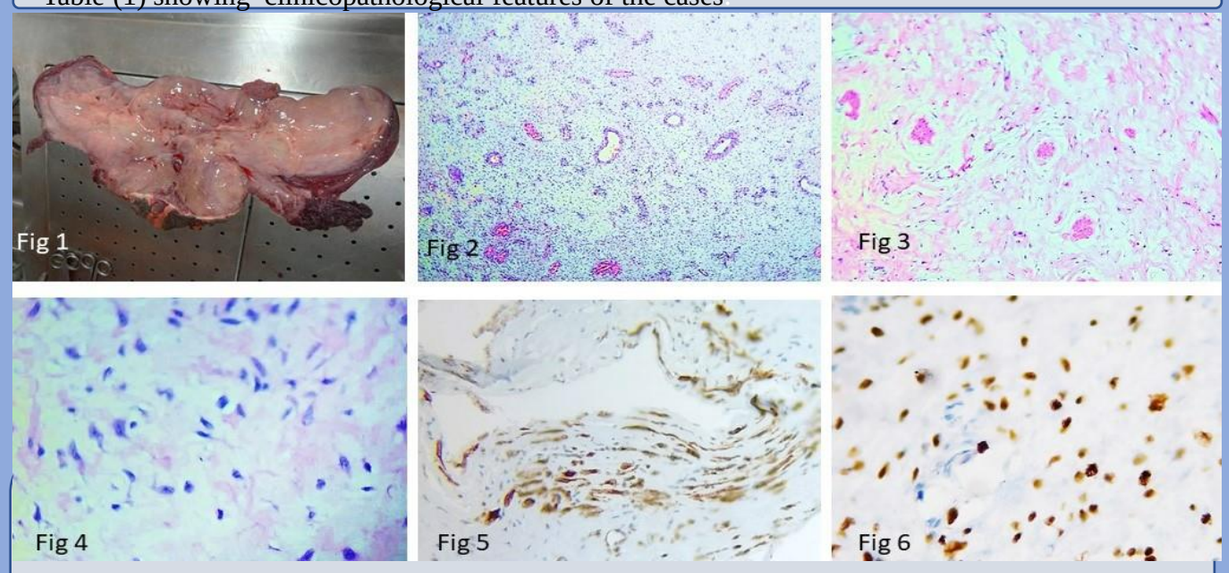

Figure (1) Cut surface is tan pink with gelatinous appearance (2) Small tumour cells embedded in myxoid to loose collagenous stroma, with varying sized blood vessels {H&E 40X} (3) Loosely organised collections of eosinophilic myoid cells {H&E 100X} (4) Small oval to spindle shaped cells with relatively bland nuclei {H&E 400X} (5) Smooth muscle actin is expressed by the perivascular myoid cells and the smooth muscle cells of the vessel wall{IHC 100X}(6) Strong and diffuse nuclear positivity for ER{IHC 400X}

Discussion and Conclusion

- AAMs typically occurs in the pelvis and perineum of reproductive aged women.
- Recognition of this entity is crucial because of its high recurrence rate (35-76%).[2,3]
- Besides the classical histomorphology as seen in our cases, uncommon features like hypercellularity, extensive collagenous deposition with or without hyalinized blood vessels or neurofibroma-like appearance can be encountered.[4]
- Presence of loosely organized collections of myoid cells situated around nerves and vessels is a characteristic finding
- Immunohistochemically, AAMs are usually positive for smooth muscle markers, ER and PR with variable CD 34 reactivity.
- Nuclear expression of HMGA2 is seen in many cases.[5]
- Differential diagnosis includes angiomyofibroblastoma, prepubertal vulval fibroma, massive vulval oedema, etc.
- Complete surgical excision is the treatment of choice.
- Administration of a gonadotropin-releasing hormone agonist is helpful.
- To conclude, we reported a group of relatively rare cases and described their clinical manifestation and pathological features.
- We also aspire others to keep this entity in mind while dealing with such vulval masses.

References

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