

A Rare Case of Deep Aggressive Angiomyxoma of the Vulva

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INTRODUCTION

The objective of this case report is to describe the clinical presentation, assessment, and management of a large vulval mass diagnosed as Deep Aggressive Angiomyxoma (DAA). DAA is a rare, infiltrative spindle cell soft tissue tumour that can arise in the lower genital tract, perineum, and pelvis, with high local recurrence rate. This report aims to raise awareness about the clinical challenges in diagnosing DAA and highlights its implications for patient management.

DISCUSSION

DAA is a rare but important differential diagnosis for vulval masses, especially in women of reproductive age. Given its potential high local recurrence rate, and its challenging nature in diagnosis, awareness of this entity is crucial for clinicians.

Early recognition through imaging and biopsy is essential for distinguishing DAA from other vulval masses, such as lipomas, schwannomas, Bartholin cyst, and Gartner duct cyst. Although DAA is benign, its aggressive local behaviour and tendency to recur necessitates a multidisciplinary approach to management, involving gynaecologists, pathologists, and oncologists.¹ Certain factors, such as incomplete surgical margins and failure to isolate the tumour, increases disease recurrence rate by 30% up to the next 15 years.¹ In rare cases, distant metastasis can also occur, further worsening prognosis. Accordingly, this case highlights the importance of long-term follow-up in preventing disease recurrence and optimising appropriate treatment.

CONCLUSIONS

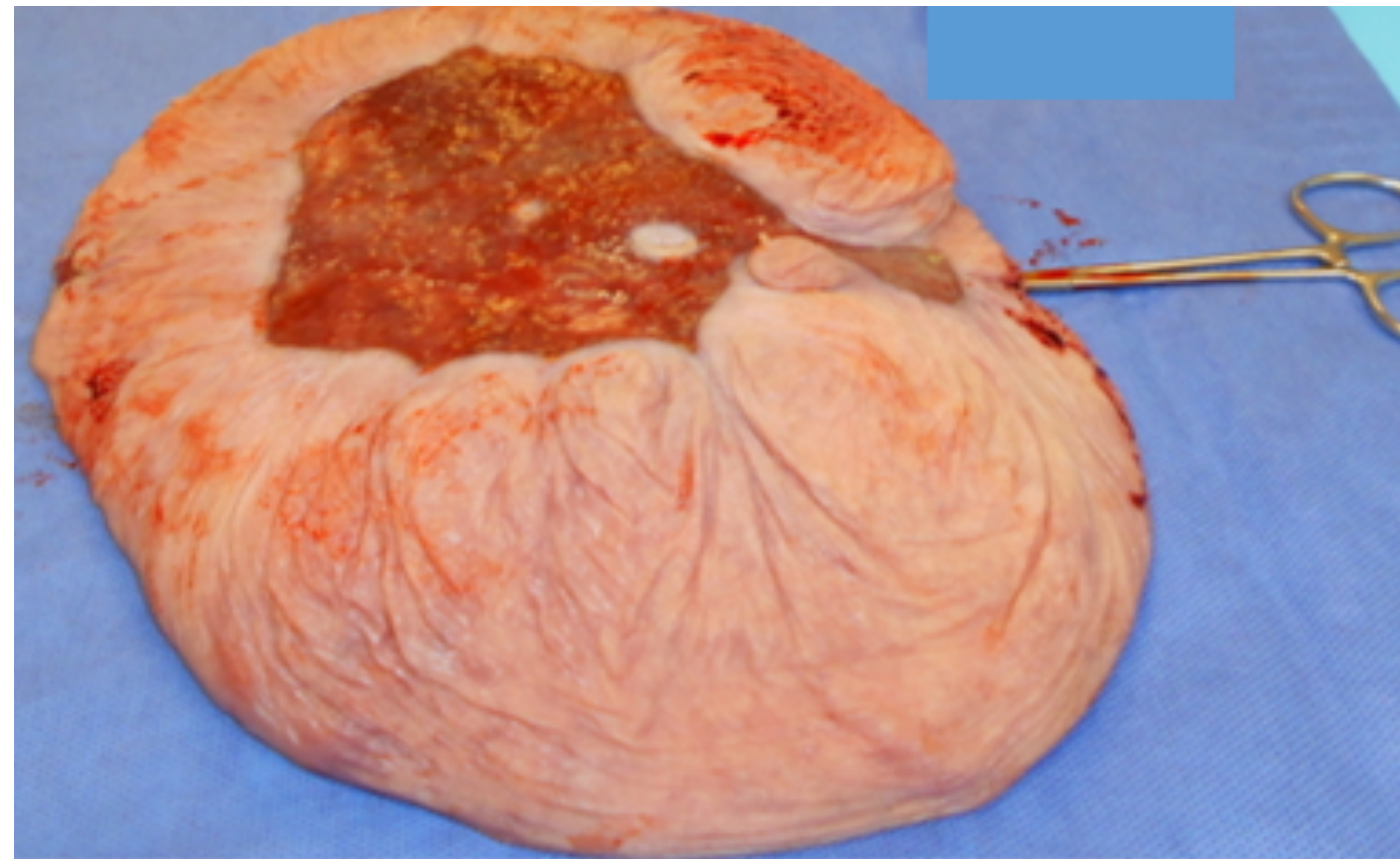
This case report emphasizes the need for vigilance in the diagnosis of rare vulval tumours like deep aggressive angiomyxoma. Surgical excision is the treatment of choice. However, due to its tendency for recurrence, long-term follow-up is recommended. Clinicians should consider DAA in the differential diagnoses of vulval masses, particularly when the mass is large or presents with unusual clinical features, to guide appropriate management and improve patient outcomes.

CASE REPORT

The patient is an 18 year old female, who presented with a large, painless vulval mass arising from the labia majora. She stated that it started similar to a skin tag, and grew over the period of one year, and more significantly so over the two months prior to her attendance. On clinical examination, this was a pedunculated, skin covered mass attached to the left labia majora, 25.5x21x5.5 cm in size, with an overlying ulcerated area of 9x8cm. It was soft and non-tender.

Surgical excision was performed, and histopathological analysis revealed the mass to be consistent with Deep Aggressive Angiomyxoma, ill-defined tumour edges extending to the subcutis. The report concludes that excision is probably complete. The patient had an uneventful postoperative recovery. She had a small stalk which was removed at a later date. Histopathology was negative for recurrence.

Aggressive angiomyxoma is an infiltrative spindle cell neoplasm arising in the soft tissues of the lower genital tract, perineum and pelvis. Local recurrence rate is approximately 30%-40%, but distant metastasis and death from disease are exceptionally rare. In some occasion recurrence may be decades after excision.



REFERENCES

¹*Aggressive angiomyxoma-vulva* (12 November 2020) *Pathology Outlines - Aggressive angiomyxoma-vulva*. Available at: <https://www.pathologyoutlines.com/topic/vulvaaggressiveangiomyxoma.html> (Accessed: 03 June 2025).