MYXOID DERMATOFIBROSARCOMA PROTUBERANS OF THE VULVA IN A POSTNATAL WOMAN: A CASE REPORT

Archana Nagendiram1, Jonathan Sandeford2,3, Francesca Watts3,4, Pranav Dorwal5,6, Sarah Lonie7, Selvan Pather2,3

Background: Vulval Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous sarcoma that develops from the dermis, with less than 70 reported cases. This case discusses vulval DFSP in a postpartum woman.

Aims: To discuss a holistic approach to vulval DFSP - with the balance of surgical excision against functional, psychosexual and cosmetic outcomes.

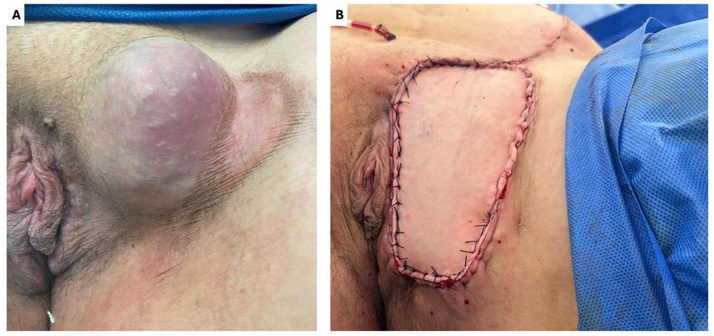

Figure 1: Pre- and post-operative images

Case: A 24-year-old woman noticed a rapidly enlarging vulval mass during pregnancy, which was an 8cm left sided vulval mass which stretched from the periclitoral area to the inguinal crease. A core biopsy was showed an indeterminate low grade myxoid lesion. Postpartum, she was referred to a Gynaecological Oncologist at a tertiary centre where it was discussed with Sarcoma and Plastic Surgeons. A radical local excision of vulvar mass, mesh reconstruction of rectus sheath and left inguinal ligaments and superficial circumflex iliac perforator flap of the left thigh was performed.

Results: Initial histopathology exhibited a myxoid lesion with negative staining including CD34, which is characteristic for DFSP. Due to indeterminate histopathology, RNA-sequencing was used and showed disease-defining chromosomal fusion (COL1A1::PDGFB fusion), which assisted the diagnosis, as this is present in over 90% of tumours. The tumour was excised over 10mm laterally and 5mm deep.

Conclusion: Over 70% of DFSP is positive for CD34 and this shows the importance of RNA-sequencing in cases of indeterminate histology. Due to high recurrence, guidelines have suggested lateral margins from 10mm up to 30mm. Though, in a young woman this was considered against long-term sexual and functional outcomes. She will undergo close surveillance both clinically and with MRI.

1 DEPARTMENT OF OBSTETRICS AND GYNAECOLOGY, ROYAL PRINCE ALFRED HOSPITAL
2 DEPARTMENT OF GYNAECOLOGY ONCOLOGY, CHRIS O'BRIEN LIFEHOUSE
3 FACULTY OF MEDICINE AND HEALTH, THE UNIVERSITY OF SYDNEY CENTRAL CLINICAL SCHOOL
4 DEPARTMENT OF TISSUE PATHOLOGY AND DIAGNOSTIC ONCOLOGY, ROYAL PRINCE ALFRED HOSPITAL
5 DEPARTMENT OF PATHOLOGY, MONASH HEALTH
6 SCHOOL OF CLINICAL SCIENCES, MONASH UNIVERSITY
7 DEPARTMENT OF PLASTIC SURGERY, ROYAL PRINCE ALFRED HOSPITAL

Chris O'Brien Lifehouse

ROYAL PRINCE ALFRED HOSPITAL