

A Rare Pathology, Vulval Leiomyoma: Case Report and Literature Review

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Abstract: This is a case of a vulval leiomyoma in a 50-year-old parous patient. There are fewer than a hundred and twenty cases of vulval smooth muscle tumour reported in literature. This case offers more evidence for this rarely reported condition, and as a learning point to avoid misdiagnosis.

Keywords: Vulval swelling, Bartholin's cyst, Marsupialisation, Uterine leiomyomas.

CASE PRESENTATION

A fifty-year-old parous woman was seen in the gynaecology outpatient department. She presented with an eight-month history of left sided vulval swelling. There had been no increase in size in recent months, the swelling was painless and did not interfere with her day-to-day life. Her cervical smears were up to date and normal. Her past surgical history included bilateral salpingo-oophorectomy and a right hemicolectomy for ileocaecal volvulus. Vaginal examination revealed a 3x2 cm firm, smooth and non-tender swelling with a cystic central aspect, in left labia minora in the typical location of the Bartholin's glands. There was no inguinal lymphadenopathy. The clinical impression was that of a Bartholin's cyst and she was placed on the waiting list for marsupialisation.

At operation, a longitudinal incision was made at most fluctuant point. There was no discharge; rather the gland appeared thickened with sheets of hyperplastic tissue. The whole gland was enucleated and the dead space closed with interrupted vicryl sutures. The overall surgical impression was that of hypertrophic Bartholin's gland. Macroscopic histology showed two pieces of nodular pale tissue, the larger 3 x 4 x 2.5cm and the smaller up to 1.3cm across. Microscopy revealed a solid spindle shaped nodule which stained with smooth muscle desmin and actin. The features were consistent with a leiomyoma.

DISCUSSION

Leiomyomas are benign soft tissue tumours that arise from smooth muscle and account for

approximately 3.8% of all benign soft tissue tumours [1]. They can develop anywhere in the body where smooth muscle is present, the most common site being the uterine myometrium. Uterine leiomyomas, more commonly referred to as fibroids, are the most common pelvic tumour in women [1].

In our case the correct diagnosis was missed prior to surgery. Given histopathology results were consistent with benign leiomyoma, the patient avoided further surgery and the outcome was good. Had the assessing clinician noted any atypical appearances of the vulva in clinic they may have considered an initial biopsy of the lump, to rule out malignancy prior to excision.

Fewer than 120 cases of smooth muscle tumours of the vulva have been reported in the literature [2]. Owing to the small amount of reported cases it is difficult to get accurate data on the various histopathologies. In 2002 Kajiwara *et al* performed a literature review in 2002 which showed in a selection of 36 cases of vulval smooth muscle tumours (2 of which were their own cases); 27 (75%) were typical leiomyoma, 4 were atypical leiomyoma and 5 were leiomyosarcoma [3]. In our case the histopathology report showed a typical leiomyoma.

Extrauterine leiomyomas are uncommon. The rarity of these tumours can present a diagnostic challenge. Levy *et al.* presented two cases of vulval smooth muscle tumour in 2014, both times initially diagnosed as Bartholin's cyst. Our patient was also erroneously diagnosed with the same prior to surgery [4]. They suggest biopsy may be advisable prior to theatre if the cyst appears firm or there are any atypical features to the history. Leiomyomas will be firm unless there is some change. Bartholins cysts will always be cystic and are located at 5 and 7 o'clock behind the labia

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majora. Correct diagnosis prior to surgery is more critical in the finding of a leiomyosarcoma, which has malignant potential. There are several case reports in the literature of vulval leiomyosarcoma presenting in a similar manner to our patient.

Preoperative figure may help clarify the diagnosis of these cases. Vulva leiomyomas typically have a characteristic finding of low signal intensity mimicking that of a smooth muscle on T2-weighted figures. This has been described as the key to diagnosis. The MRI signal in these tumours is isotense to that of muscle on the T1-weighted figures, and the tumours enhance homogeneously after the administration of contrast material [5].

Leiomyoma are benign monoclonal tumours arising from smooth muscle. Although there are reported leiomyoma variants that may display some malignant features, these are extremely rare, and leiomyoma is considered a benign condition [1]. The diagnosis of extrauterine leiomyosarcoma in the gynaecological tract requires the presence of at least three of the following characteristics; infiltrative margins, diameter >5cm, >5 mitotic figures per 10 high power field, and moderate to high cytological atypia. Lesions with one of the above are diagnosed as leiomyomas; whereas cases with two of the above characteristics are labelled as atypical leiomyomas [6].

Whilst the treatment practices in the UK for uterine leiomyoma are well established, there is limited guidance around treatment and follow up for extrauterine leiomyoma. Neilsen *et al* performed a clinopathological study of 25 cases of vulval smooth muscle tumours and reviewed the literature. Most of these swelling were diagnosed as Bartholin's cyst prior to surgery. There were 20 cases with leiomyoma (4 atypical) and 5 with leiomyosarcoma. There was recurrence of disease in 1 of 20, and 4 of 5 cases with leiomyoma and leiomyosarcoma respectively. This raises the debate and perhaps relevance of follow up even in cases of leiomyomas, which are benign. They advised leiomyomas should be excised conservatively, whilst sarcomas require wide excision with negative margins [6]. Given the different surgical techniques, this may support the practice of biopsy prior to excision of the lesion, in order to plan ahead for the surgery.

HISTOLOGY PICTURES

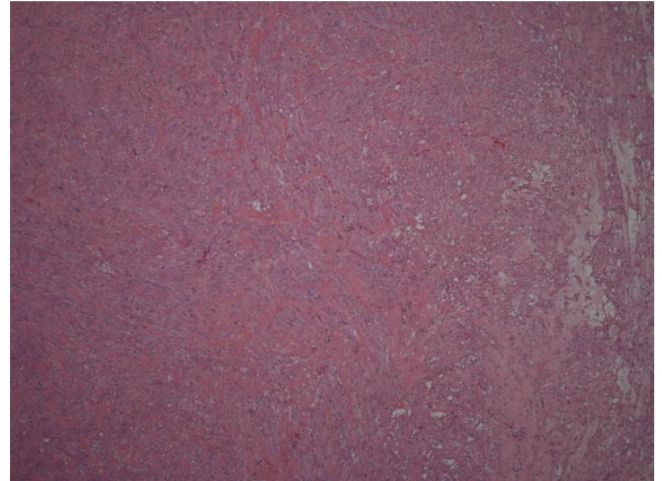


Figure 1: Low power view showing a benign spindle cell neoplasm with some areas of degenerative changes.

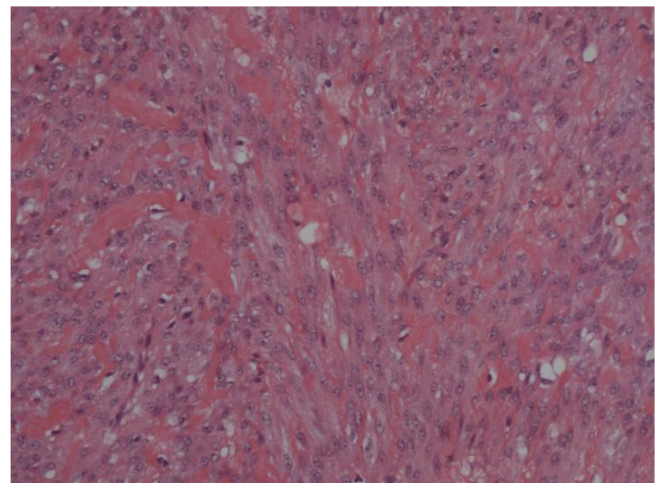


Figure 2: High power view showing bland spindle cells with intervening collagenous stroma.

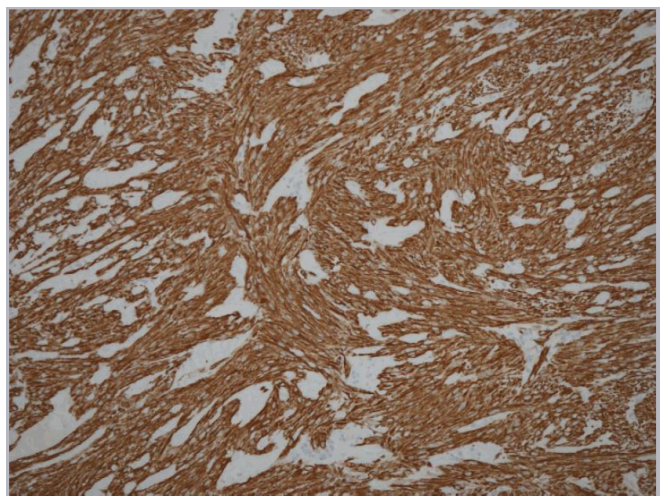


Figure 3: Immunohistology showing strongly positive for desmin.

LEARNING POINTS

We present this rare and interesting case. Evidence is limited with regard to treatment and follow-up of this condition. It serves as a good reminder to keep an open mind as to differential diagnoses of vulval swelling, which is a common presentation to gynaecology outpatient departments and wards.

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