Labial adhesion resulting from pemphigus vulgaris: A rare cause of hematometra

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INTRODUCTION:

- Pemphigus vulgaris (PV) incidence: 0.76- 16.1 cases/1,00,000 individuals per year
- Autoimmune disease: desmoglein-1 & 3 protein
- Organs Involved: mucosa of oral cavity (M/C), oesophagus, larynx, nose, nails, genital tract
- Diagnosis: Clinical features, histology, immunofluorescence (IF)
- Treatment: steroids, immunosuppressants
- Here, we describe labial adhesion in reproductive age group female arising out of PV leading to secondary amenorrhea and hematometra
- Never been reported before

CASE PRESENTATION:

- A female in her 20's, diagnosed case of PV presented with amenorrhea and cyclical abdominal pain for 2 months
- No urinary/bowel complaints
- No sexual trauma/ assault
- Past menstrual history: unremarkable with menarche at 15 years
- Past history: Onset of oral, nasal and genital erosions 8 months back, leading to difficulty in eating solids & liquids. Diagnosed with PV after histology and IF from oral mucosa biopsy
- Treatment received: injection dexamethasone pulse therapy, tablet prednisolone 30 mg daily & tablet azathioprine 50 mg twice daily. Genital lesions had resolved at presentation. Oral and nasal lesions were in the healing phase.
- Family history: Not significant

Examination:

General Physical Examination:

Crusted erosions on both the lips, hard and soft palate, nasal mucosa.

Normal secondary sexual characteristics

Abdomen: soft, non-tender, suprapubic bulge corresponding to 12 weeks pregnant uterus size appreciated

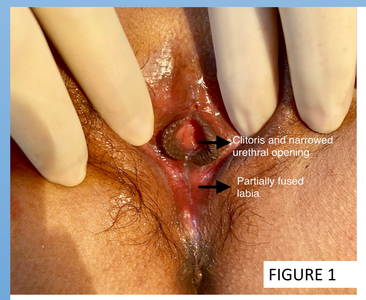
Genital Examination

Thick adhesion band between labia minora obliterating the vaginal introitus with only 1 cm unfused area anteriorly.

Clitoris visible and urethral orifice narrowed

No erosions or erythema visualized in the genital area (Figure 1)

Per-rectal examination: Same mass felt anteriorly



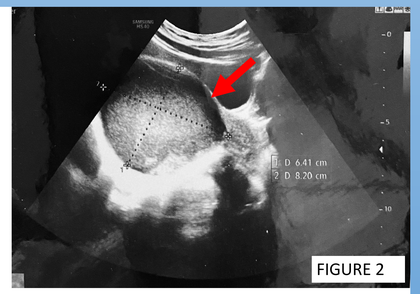


Figure 1: External genital examination showing partially fused labia, clitoris and narrowed urethral opening.

Figure 2: Trans-abdominal longitudinal ultrasound image revealing hematocolpos measuring 6.4X8.2cm (Red arrow)

INVESTIGATIONS:

- Routine tests- unremarkable
- Abdominal ultrasonogram (US)- hyperechoic collection in cervix -6.4 X 8.2 cm (hematocolpos) and in uterus 4.5 x3 X 2.3 cm (hematometra)
- Tzanck smear from labia- acantholytic cells, neutrophils

TREATMENT:

- Multidisciplinary approach (Gynaecologist, dermatologist)
- Plan: Topical triamcinolone acetonide twice daily (0.1%) followed by surgical adhesiolysis in view of thick nature of fusion band
- On day 2 of triamcinolone application, she started to bleed through vagina
- On examination: labia were still fused, adhesion band had thinned out, bleeding seen coming through vagina
- Repeat US: Empty uterine cavity and cervix
- Surgical adhesiolysis cancelled. Manual separation of labial adhesions under local anaesthesia successfully attempted. Cervical opening felt. Blood observed draining. No erosions were visualized (Figure 3 & 4).

FOLLOW-UP:

Topical triamcinolone acetonide continued twice daily for 1 week followed by OD for 3 wks. At the end of 1 year follow up, labia were still separated, no recurrence.

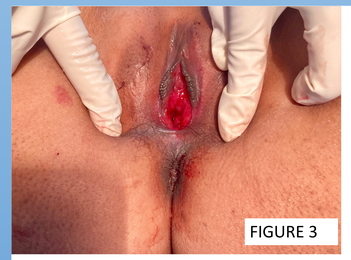


Figure 3: Genital area after topical steroid application and spontaneous onset of vaginal bleeding

Figure 4: Post manual lysis of the adhesion band. Genital area depicting unfused labia minora and canalized vagina smeared with blood.

DISCUSSION:

- Atypical presentations of genital pemphigus vulgaris described in literature include desquamative vaginitis and squamous cell carcinoma but labial adhesion and hematometra have not been reported.
- Also, labial fusion is a rare clinical entity in reproductive age group and cause is unknown. Current case: could be due to genital area inflammation and superadded infection
- In the current case, less possible causes include mucus membrane pemphigoid, erosive lichen planus and lichen sclerosus.
- No guideline available for labial adhesion but few studies have recommended topical estrogen or steroid application for 4-6 weeks in prepubertal girls and surgical therapy in reproductive women as first line.

CONCLUSION

- Ulcerative mucocutaneous lesions like pemphigus vulgaris should be kept as a differential diagnosis of labial adhesion in relevant clinical scenario.
- Two-step procedure (topical therapy followed by manual separation of adhesion band) can be tried with thick adhesion band as it will avoid surgery related risk.

References:

1. Akhyani M et al. Cervicovaginal involvement in pemphigus vulgaris: a clinical study of 77 cases. Br J Dermatol. 2008 Mar;158(3):478–82.