***no patient handout*

Fox-Fordyce disease

Synopsis

Fox-Fordyce disease is a rare inflammatory condition of apocrine gland-bearing regions caused by obliteration of the follicular infundibulum with keratin. The etiology is unknown, although an endocrine role has been postulated. The disease manifests as intensely pruritic skin-colored or keratotic papules in the axillae, pubic, and periareolar areas. Heat, humidity, and stress may be exacerbating factors.

It is most common in females during adolescence, but it can be seen between puberty and age 35. Less commonly, it can affect male patients.

Codes

ICD10CM:
L75.2 – Apocrine miliaria

SNOMEDCT:
65038009 – Fox-Fordyce disease

Look For

Multiple monomorphous, small (2-3 mm) white to skin-colored to light-brown to yellow papules in apocrine-dense areas such as the axillae, areolae, and pubic areas (mons pubis and [in women] labia majora).

Each papule is perifollicular and has a central punctum. Keratotic material may be expressed. Sometimes the papules may contain a milky fluid.

Axillary and pubic hair may be sparse, and there may be associated excoriations and/or lichenification from scratching.

Diagnostic Pearls

The pruritus is extreme and is out of proportion to the lack of redness. Fractured hair shafts are commonly seen overlying the papules as a result of itching and scratching.

Differential Diagnosis & Pitfalls

- **Folliculitis**
- **Acanthosis nigricans**
- **Lichen simplex chronicus**
- **Syringomas**

- **Intertrigo**
- **Candidiasis**
- **Epidermal nevus**
- **Follicular eczema**
- **Molluscum contagiosum**
- **Flat warts** / **genital warts**
- **Lichen nitidus**
- **Milia**
- **Miliaria rubra**
- Adnexal tumor
- **Trichostasis spinulosa**

Best Tests

This diagnosis can often be made on clinical grounds, but skin biopsy will be confirmatory. Skin biopsy demonstrates obstruction of apocrine ducts with an inflammatory infiltrate.

Management Pearls

Palliative topical antipruritic formulations such as pramoxine with menthol and camphor may be useful adjuncts.

In female patients, oral contraceptives (or other alterations of the hormonal milieu, eg, continuous progesterone, spontaneous menopause) can be a useful adjuvant to therapy.

Exercise and other activities that lead to sweating may exacerbate the condition.

Therapy

Treatment can be difficult and complicated by skin irritation. Treatments that have been successful include the following.

Intralesional (triamcinolone) and mid-potency topical corticosteroids (class 3-4):

- Triamcinolone cream, ointment – Apply every 12 hours (15, 30, 60, 120, 240 g), or
- Mometasone cream, ointment – Apply every 12 hours (15, 45 g), or
- Fluocinolone cream, ointment – Apply every 12 hours (15, 30, 60 g).

Topical retinoids:

- Tretinoin 0.025% applied nightly.

Oral contraceptives have demonstrated efficacy in some women.

Topical clindamycin (Cleocin-T) applied to affected areas twice daily.

Surgical excision is an option for severe cases. Liposuction to remove the apocrine glands may offer relief without the morbidity of excision.

Drug Reaction Data

Below is a list of drugs with literature evidence indicating an adverse association with this diagnosis. The list is continually updated through ongoing research and new medication approvals. Click on Citations to sort by number of citations or click on Medication to sort the medications alphabetically.

Medication	Citations
Estrogen	2
Oral contraceptives	2