Case 5 : Follicular Occlusion Triad (FOT)

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CASE SUMMARY

History

Mr. Ho is a 38-year old electrician. He presented with a two weeks' history of acute painful erosions and ulcerations over the scalp, face and buttock. There were severe purulent discharges from these sites and the patient complained of a mild fever. He was admitted into Tuen Mun Hospital under the care of the general surgeon. Multiple wound swabs were taken but the results were all insignificant. Parenteral antibiotics of ampicillin and cloxacillin were given but the patient did not respond. Dermatological opinion was consulted. He had no past or family history of severe acne.

Physical examination

The scalp showed multiple, large fluctuant discharging abscesses, clinically resembling dissecting cellulitis (Figure 1). The buttock was covered with some ulcerated annular plaques with the largest measured 3 cm in diameter (Figure 2). There were severe nodulocystic acnes on the face (Figure 3).

Investigation

The baseline haematology and biochemistry were normal and swabs for culture from all discharging sites were negative. The fungal culture from the scalp was negative.

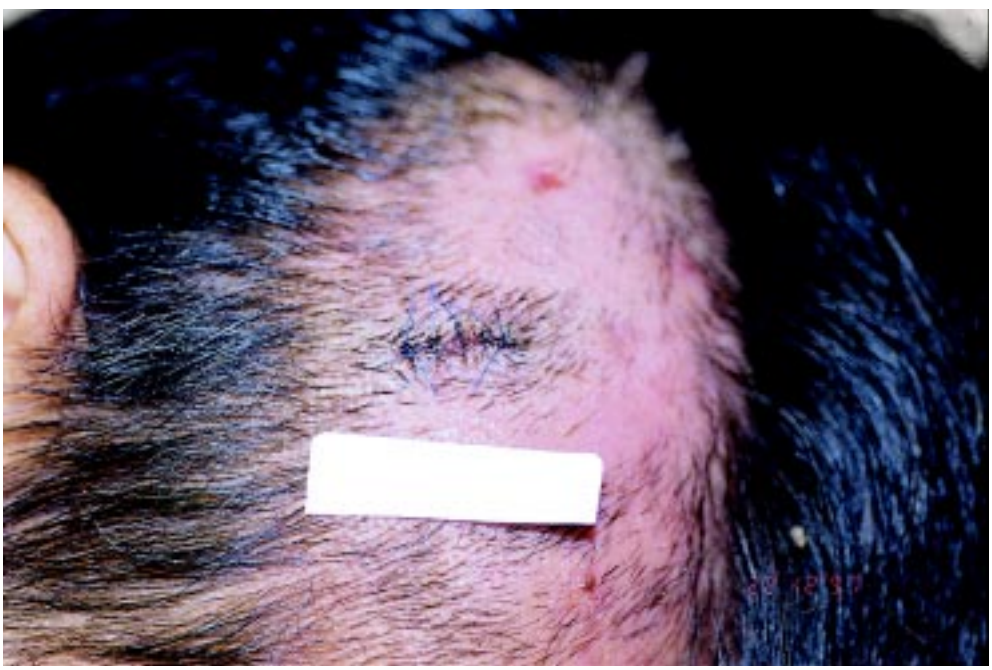

Figure 1: Multiple large, 1-2 cm, mobile fluctuant abscess over the scalp.

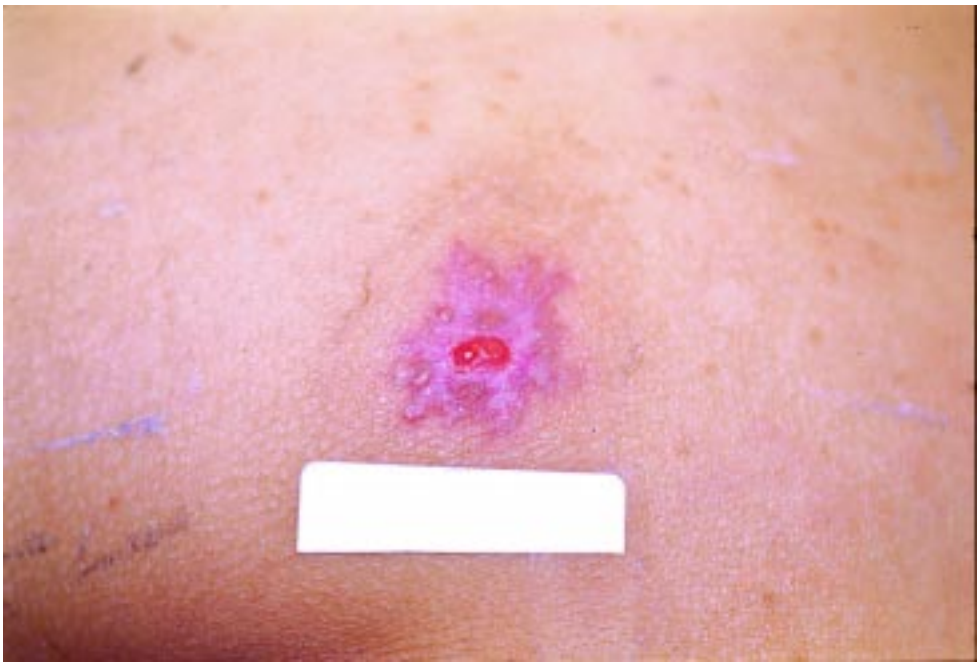

Figure 2: Large 3 cm in diameter purulent discharging abscess over the buttock.

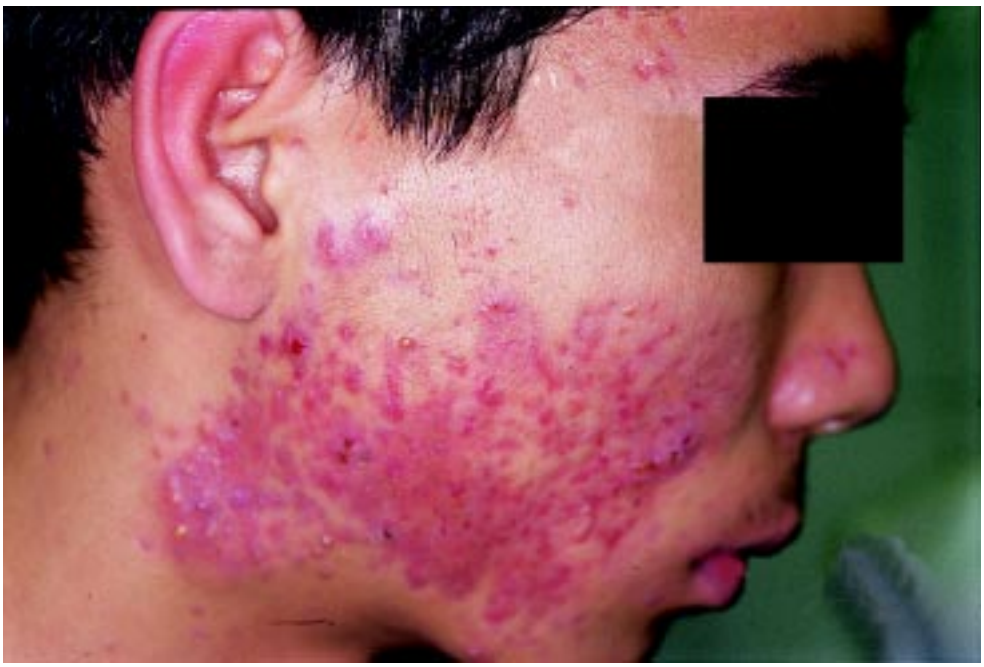

Figure 3: Severe nodulocystic acne vulgaris.

An incisional biopsy of the scalp was performed. It showed a marked perifollicular infiltrate of neutrophils, lymphocytes and histiocytes with disruption of the pilosebaceous structures and abscess formation. The infiltrate extended into the panniculus. Follicular hyperkeratosis with plugging was noted in the nearby follicles. Fungal stain was negative. The diagnosis was acute folliculitis of the scalp consistent with Follicular Occlusion Triad.

Management

The patient was subsequently started on Minocycline MR 100 mg daily. He showed marked improvement with desiccation and shrinkage of the ulcers. Unfortunately, the affected areas showed marked post inflammatory hyperpigmentation and scarring. He was followed up regularly to observe for any relapse.

REVIEW ON FOLLICULAR OCCLUSION TRIAD (FOT)

It is a rare cutaneous disorder referring to the association of dissecting cellulitis of the scalp, acne conglobata and hidradenitis suppurativa occurring in the same patient. The condition has in common the occlusion of the sebaceous or apocrine glands with subsequent inflammation and scarring.

Follicular occlusion tetrad refers to the same spectrum of disease with the addition of pilonidal sinus in the same patient.

It was first reported by Brusting in 1952[1]. Self and Montes coined the term follicular occlusion triad in 1970[1]. In 1982, Rosner conducted a retrospective study involving 10 patients with hidradenitis suppurativa and dissecting cellulitis of the scalp[1]. He found only 3 of the cases had FOT. There is no recent review or study found in the dermatological literature on this rare and interesting condition. The last detailed illustration was reported in the Journal of Rheumatic Disease Clinics of North America in 1992.

Clinical features

Acne conglobata (AC) is a severe form of acne vulgaris characterised by suppurative cystic lesions on the face with interconnecting sinuses. The lesion can develop in other sites like the buttocks, thighs, upper arms, neck and the trunk. Scarring and post inflammatory pigmentary changes are common. It commonly affects men and tends to follow a chronic and unremitting course[2].

Hidradenitis suppurativa (HS) is a chronic inflammatory condition of the skin characterised by suppurative nodules, cysts, abscesses and sinus tracts of the cutaneous tissues. The common sites of involvement are the axillae, groin, and the perianal area. In the past, it was believed to be due to the occlusion of the apocrine glands. Disfiguring scars are common. A chronic course is the rule. However, it is said to be more common in middle aged female[3].

Dissecting cellulitis of the scalp (DC) is a rare disease of the scalp. It is also known as **perifolliculitis capitis abscedens et suffodiens**. It is characterized by tender suppurating nodules over the scalp with intercommunicating sinuses. The nodules or abscesses are usually fluctuant and healed with scarring alopecia. It usually occurs in male, black patients and sometimes a positive family history can be elicited. The disease tends to be chronic and is difficult to treat[4].

Follicular occlusion triad (tetrad) (FOT) is a combination of the above three conditions occurring in the same patient. When pilonidal sinus or cyst is also present, this is referred to as follicular occlusion tetrad. It is a very rare disease and is not well studied clinically and epidemiologically. Patients with FOT in the United States are usually black and lack constitutional symptoms[1].

Laboratory investigations

Complete blood picture shows normal or mildly elevated white cell count and mild anaemia. Erythrocyte sedimentation rate is elevated at 30 - 70 mm/hr. Rheumatoid factor, antinuclear antibody, serum complement levels are within normal limit. Circulating immune complexes are occasionally found. Serum protein electrophoresis occasionally demonstrates alpha or beta hypergammaglobulinaemia. Occasionally elevated Ig A is present. HLA - B 27 is negative. Microbiological cultures are not helpful apart from growing secondary pathogens.

Skin biopsy provides only ancillary support. The diagnosis mainly depends on the clinical features.

Summary of clinical and epidemiological findings of the above conditions:

	HS[3]	AC[2]	DC[4,5]	FOT[1]
Age range	17-59	18-30	18-40	22-46
Sex ratio F:M	8:1	M>F	3:1[5]	exclusively male
Ethnicity	no racial differences	no racial differences	Afro-Caribbean	Black
Morphology	abscesses, ulceration, sinus tracts, scarring	burrowing, abscesses, scarring	fluctuant abscesses, cysts, sinus, patchy hair loss, scarring alopecia	HS + AC + DC
Distribution	axilla, groin, perineum, buttock	face, neck, trunk, buttock	scalp, beard, pubic area	scalp, face, axilla, buttock
Course	chronic	chronic	refractory	exacerbation/ remissions
Site of lesion	apocrine gland	sebaceous glands	hair follicle	follicular apparatus
Complication	scarring, squamous cell carcinoma	scarring, keloid, squamous cell carcinoma	scarring, alopecia, metastasizing squamous cell carcinoma	as in HS, DC, AC
Treatment	difficult	systemic retinoids	difficult	Minocycline
Association	obesity, DM, arthritis, DC, AC	arthritis, DC, HS	HS, AC	arthritis
HLA status	DRw4 if associated with AC	DRw4 if associated with HS	unknown	unknown

Pathogenesis

HS, DS, AC and FOT are believed to have a common pathogenic basis[1]. Occlusion of the follicular apparatus seems to play a primary role. The poral occlusion may involve both apocrine and sebaceous glands. The active inflammation is likely to occur in the apocrine glands in HS and sebaceous glands in AC.

Genetic factor may play a role as suggested by the fact that isolated cases of DC and AC developed in identical twins. Furthermore, in HS, some patients show a positive family history. However, HLA tissue typing is not conclusive although all patients with both HS and AC possess DRw4 status[1].

Infection did not seem to play a central role in the pathogenesis of the conditions.

Immune mechanism has been postulated by some authors as the main pathogenesis. Circulating immune complexes formed by type III reaction has been found in a study of 13 patients. These patients were also found to have a low C3 level and raised gammaglobulins. The antibodies involved were mainly the Ig A and Ig M subtypes[1]. Immunomodulating drugs like systemic corticosteroids and isotretinoin were reported to be effective in alleviating these conditions.

Learning points:
The diagnosis of FOT mainly depends on clinical features and the condition should be suspected in patients who have a history of severe acne, develop refractory discharging abscesses, sinuses and ulcerations at scalp, axillae, groin and buttocks.

Summary of treatment modalities of the 4 conditions are listed below:

	HS[3]	AC[2]	DC[4,5]	FOT[1]
Topical therapy	no response	no response	Vaseline	no response
Systemic antibiotics	no response	high dose tetracycline or erythromycin	poor response	Minocycline
Systemic steroid	no response	intra-lesional triamcinolone	poor response	no response
Systemic isotretinoin	good response (0.71- 1.2 mg/ kg/day for 4 to 6 months)	treatment of choice (1 mg/ kg/day for 4 to 6 months)	good response (0.75- 1 mg/ kg/day for 9 to 11 months)	theoretical good response (no data on dosage/duration)
Other suggested systemic therapy	anti-androgen cyproterone acetate, Eugynon 50, spironolactone	Dapsone	Zinc sulphate 300 mg T.D.S. for 3 months (successful in isolated case)	not known
Surgery	wide excision with grafting	extraction of comedones	excision with grafting	as in HS,AC,DC
X ray	not mentioned	not mentioned	X ray epilation	not mentioned
Laser therapy	not mentioned	not mentioned	CO2 laser	not mentioned

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