

A Very Rare Case of Dissecting Cellulitis of the Scalp in an Indonesian Man

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Abstract: Dissecting cellulitis of the scalp (DCS), also known as dissecting folliculitis, perifolliculitis capitis abscedens et suffodiens (PCAS), or Hoffman's disease, is a primary neutrophilic cicatricial alopecia without clear etiology. Along with hidradenitis suppurativa, acne conglobata, and pilonidal cyst, they were recognized as 'follicular occlusion tetrad'. A 43-year-old Indonesian man presented to our department with four years history of persistent, slightly painful subcutaneous nodules, abscesses, and sinuses that discharged purulent exudate on vertex and occipital scalp. There was also associated patchy alopecia. He had severe acne during his adolescence to early adulthood. Trichoscopic evaluation showed yellowish and whitish area lacking of follicular openings. Histopathological examination showed follicular occlusion, dilatation, and rupture with mixed inflammatory infiltrates, mainly neutrophils. The diagnosis of DCS was confirmed by clinical, trichoscopic, and histopathological examinations. Isotretinoin 20 mg daily was given to normalize the follicular keratinization. Considering its very rare occurrence in an Indonesia man, this case was reported to emphasize the diagnosis of DCS.

1 INTRODUCTION

DCS, also known as dissecting folliculitis, PCAS, or Hoffmann's disease, is a very rare primary neutrophilic cicatricial alopecia without clear etiology (Badaoui et al., 2016). It was first described by Spitzer in 1903 who termed the disease "dermatitis follicularis capitis et perifollicularis conglobata" (Spitzer, 1903). Hoffman then termed it as PCAS in 1908 (Hoffman, 1908). Since the first description to 2014, the details of only 72 patients have been published. DCS has been considered to be a part of 'follicular occlusion triad', along with hidradenitis suppurativa and acne conglobata (Otberg & Shapiro, 2012). Other literature included pilonidal cyst and altogether they were recognized as 'follicular occlusion tetrad' (Badaoui et al., 2016). Its chronic relapsing courses resulted in cicatricial alopecia with hypertrophic or keloidal scars formation (Otberg & Shapiro, 2012). Various treatments, such as systemic antibiotics, intralesional corticosteroid, oral prednisolone, and isotretinoin showed clinical improvement (Otberg & Shapiro, 2012; Scheinfeld, 2014). In refractory and more advanced cases, anti tumor necrosis factor alpha (anti-TNF α) and surgery

should be considered (Otberg & Shapiro, 2012; Scheinfeld, 2014). Considering its low prevalence in Indonesia, we are intrigued to report a case emphasizing the diagnosis of DCS.

2 CASE

A 43-year-old Indonesian man presented to our department with four years history of persistent, slightly painful lumps that discharged purulent material during compression on his vertex and occipital scalp. There was also associated patchy alopecia. Those lumps firstly appeared as small red bumps resembling folliculitis, which then enlarged. He experienced severe facial acne during his adolescence to early adulthood. He regularly washed his hair daily and got his hair cut with scissors monthly. He often wears a hat, which was washed once every one or a couple of weeks. There was no familial history with the same complaints and previous mechanical trauma. On physical examination, there were multiple flesh-colored subcutaneous nodules that fluctuated, sinuses that discharged purulent exudate, and patchy alopecia.

These lesions were slightly painful (VAS 2-3). The surrounding tissues were neither erythematous nor edematous. We also did not observe any tufted hairs. Trichoscopic evaluation showed yellowish and whitish area lacking of follicular openings. Microscopic examination with KOH 20% from the alopecia patch revealed no fungal elements. Bacterial culture from the discharge and skin tissues were done and showed growth of *Staphylococcus epidermidis*, which was still sensitive to various antibiotics. Histopathological examination showed follicular occlusion, dilatation, and rupture with mixed inflammatory infiltrates, mainly neutrophils. Based on the clinical, trichoscopic, and histopathological examinations, the diagnosis of DCS was confirmed. Isotretinoin 20 mg daily was then initiated.



Figure 1. Clinical lesions: A: Fluctuating flesh-colored nodules in vertex-occipital scalp area; B: draining sinuses with purulent discharge.



Figure 2. Trichoscopic and histological examination. A: Yellowish and whitish are lacking of follicular openings; B: Follicular occlusion and dilatation (H&E40x).

3 DISCUSSION

Dissecting cellulitis of the scalp is a very rare disease characterized by inflammatory nodules, abscesses, and sinuses, which may progress into scarring alopecia (Segurado-Miravalles et al., 2017). Along with hidradenitis suppurativa, acne conglobata, and pilonidal cyst, DCS forms the follicular occlusion tetrad (Badaoui et al., 2016; Segurado-Miravalles et al., 2017). These diseases shared similar etiopathogenesis involving hyperkeratosis, follicular occlusion, and subsequent inflammation (Segurado-Miravalles et al., 2017). Not all patients presented all these four diseases (Badaoui et al., 2016). A retrospective study of 51 patients showed that 12% patients were also presented with hidradenitis suppurativa, 16% with acne conglobata, and 4% with coexisting hidradenitis suppurativa and acne conglobata (Badaoui et al., 2016). No patients were identified as having pilonidal cyst (Badaoui et al., 2016). Mechanical trauma as a predisposing factor was only found in five patients (Badaoui et al., 2016). Subjective complaints are pain or itch (Badaoui et al., 2016). Eighty six percent patients progressed into chronic disease (Badaoui et al., 2016). The observed abscesses were usually sterile (Gaopande et al., 2015). Bacterial infection can occur secondarily during the course (Gaopande et al., 2015). Microorganisms cultured in reported cases include *Pseudomonas* species, *Staphylococcus epidermidis*, *Prevotella intermedia*, *Peptostreptococcus asaccharolyticus*, and *Propionibacterium acnes* (Gaopande et al., 2015). DCS is predominantly found in African American men between 20 and 40 years (Gaopande et al., 2015). Although it rarely affected Asian, it has been reported in other Asian ethnicities (Chinese and Indian) (Gaopande et al., 2015; Qi et al., 2014). A retrospective study by Badaoui et al (Badaoui et al., 2016). showed that the mean age was 26,6 years, with wider range of age between 15-62. Gaopande et al (2016) even reported the occurrence of this disease in a 7-year-old boy. A multicenter study which was conducted in four hospitals in Spain also reported the occurrence in a 23-year-old Asian woman (Segurado-Miravalles et al., 2017). The pathophysiology of DCS remains unclear (Badaoui et al., 2016). The young age of onset, the occurrence in patients with dark phototype, and the cases of familial DCS suggest a genetic predisposition (Badaoui et al., 2016). The predominance of patients with dark phototype raises the question of the role of hair type, which is often coarse and frizzy, as well as traumatic factor, such as hair shaving (Badaoui et al., 2016). The male dominance and vertex as the predilection

area could also suggest a hormonal risk factor (Badaoui et al., 2016). Finally, commensal bacteria may play an essential role as alloantigens in the pathophysiology of DCS (Badaoui et al., 2016). The loss of immune tolerance to these alloantigens may lead to an inflammatory reaction (Badaoui et al., 2016; Scheinfeld, 2014). To the best of authors' knowledge, this is the third case of DCS in an Indonesian man. Although unpublished, Sirait (2015) reported the occurrence of DCS associated with hidradenitis suppurativa in 2015. Rahman et al. (2017) then reported a case of follicular occlusion tetrad in 2017. This patient is a 43-year-old Indonesian man with straight hairs and no associated mechanical trauma. There was also no familial history with the same complaints. The signs and symptoms of patient reported in this report are well suited with DCS despite the lack of associated predisposing factors except male gender. Other explanations supporting the diagnosis of DCS are listed below:

1. Anamnesis: lumps that discharged purulent material on the predilection areas. History of severe acne during his adolescence to early adulthood.
2. Physical examination: observed subcutaneous nodules, abscesses, and sinuses that discharged purulent exudate.
3. Trichoscopic examination: yellowish and whitish area lacking of follicular openings. These were consistent with trichoscopic findings explained by Laccarubba et al (2017).
4. Histopathological examination: follicular occlusion, dilatation, and rupture with mixed inflammatory infiltrates, mainly neutrophils. Multiple hair shaft fragments are evident in pilonidal cyst (Calonje et al., 2012). This feature is not observed, so that the histopathology is more suited with DCS than pilonidal cyst.

DCS must be distinguished with other diseases involving scalp (Vasant et al., 2014). The tendency of DCS to cause fluctuating nodules and sinus tracts helps to distinguish it from acne keloidalis nuchae (AKN) (Vasant et al., 2014). Folliculitis decalvans, another differential diagnosis, is characterized by tufted folliculitis, in which multiple hair tufts emerge from dilated follicular orifices (Vasant et al., 2014).

Various therapeutic strategies were reported successful in treating DCS, such as systemic antibiotics (minocycline, tetracycline, cloxacillin, erythromycin, cephalosporin, or clindamycin), intralesional corticosteroid, and oral prednisolone (Otberg & Shapiro, 2012). The benefits of systemic

antibiotics were considered to be their anti-inflammatory effects rather than antibacterial (Otberg & Shapiro, 2012). Isotretinoin 0,5-1 mg/kg/day has shown prolonged remission (Otberg & Shapiro, 2012). The mechanisms of action of isotretinoin are normalizing follicular keratinization and reducing the aberrant immune responses (Scheinfeld, 2014). Anti-TNF α may be used when isotretinoin fails (Scheinfeld, 2014). It can also defer the need of a surgical treatment (Scheinfeld, 2014). Incision and drainage may be done to painful and resistant nodules (Scheinfeld, 2014). Marsupialization with curettage of the cyst wall and total scalp excision followed by split-thickness skin grafting have been reported, but these surgical procedures should only be done for extreme and refractory cases (Otberg & Shapiro, 2012). Badaoui et al. (2016) reported that 78% patients receiving systemic antibiotics (doxycycline, pristinamycin, rifampicin, or a combination of several antibiotics) showed moderate improvement. However, the disease was relapsed in all of those patients after antibiotic cessation (Badaoui et al., 2016). Seventy one percent patients had received systemic retinoid and almost all (92%) showed complete remission after 3 months (Badaoui et al., 2016). Isotretinoin is considered as the first line therapeutic option for DCS (Badaoui et al., 2016; Fransisco et al., 2017). The efficacy of isotretinoin in treating DCS has been published (Marquis et al., 2017). However, the optimal dose, duration of therapy, and combination with other agents have not been fully elucidated (Marquis et al., 2017). Marquis et al.(2017) reported an excellence therapeutic response after 4 months course of isotretinoin at 0,27 mg/kg/day. Considering the possibility of developing side effects given at higher dose, this patient received isotretinoin 20 mg daily (equal to 0,25 mg/kg/day). He has been receiving isotretinoin for the past two months and showed improvement.

4 CONCLUSIONS

DCS is a very rare case in Indonesia and should be recognized as one of differential diagnosis when seeing nodules, abscesses, and sinuses on the scalp. The diagnosis can be confirmed by clinical, trichoscopic, and histopathological examinations. The therapy can be started immediately after the diagnosis has been confirmed.

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