# Alopecic and aseptic nodule of the scalp & cutis verticis gyrata: when rarities meet

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**Background:** Alopecic and aseptic nodule of the scalp (AANS) is a rare entity characterized by the presence of sterile, culture-negative nodules/cysts and areas of non-scarring alopecia.

**Methods:** We describe a case on the vertex of a 26-year-old man with a two-week history of a nodular, moderately-fluctuant, alopecic lesion. The patient also had cutis verticis gyrata (CVG).

**Results:** The aspirate culture was negative and histopathologic findings were nonspecific. We also performed a trichoscopy of the lesion. After eight weeks, the patient showed dramatic clinical response to doxycycline and a dose of intralesional corticosteroid.

**Conclusion:** This is the first report of the simultaneous occurrence of AANS and primary essential CVG. AANS is a probably under-reported cause of non-scarring alopecia with a rapid and dramatic response to non-surgical treatment and a generally good prognosis.

**Keywords:** alopecia, corticosteroid, doxycycline

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## INTRODUCTION

A 26-year-old Caucasian man presented with a one-month history of a localized area of alopecia on the vertex. Symmetric anteroposteriorly-oriented furrows and ridges were palpated over the parietal, parietotemporal, and upper occipital areas.

## CASE SYNOPSIS

The patient did not complain of pain, burning, scaling, or pruritus. Physical exam revealed a 1×2×2 cm skin-colored, dome-shaped, non-scarring, well-circumscribed, mobile, alopecic nodule in the right parietal vertex with moderate fluctuation on palpation. No tenderness, discharge, scale, or crust was found, and the pull test was negative. Another larger, non-alopecic nodule (5×4×1 cm) was found on the occipital prominence, of which the patient had no complaint. Symmetric anteroposteriorly-oriented furrows and ridges were palpated over the parietal, parietotemporal, and upper occipital areas. All other parts of the physical exam were unremarkable. The patient had no systemic manifestations, and his past medical history was not significant. He was a non-smoker with a negative drug and relevant social history. The aspirate was of citrine-yellowish color, returning negative from

bacterial/fungal cultures. The complete blood count, biochemistry panel, urinalysis, thyroid-stimulating hormone (TSH), free T4, VDRL, and viral markers for HBV, HCV, and HIV were normal. An ophthalmologic consultation was performed with normal results. Trichoscopy showed compound follicles, fine vellus hair, and yellow dots with no black dots or dystrophic hair (Figure 1). We performed an elliptical lesional biopsy. The histopathologic study revealed nonspecific changes including an acanthotic, hyperkeratotic epidermis with areas of papillomatosis. An interstitial and perifollicular lymphohistiocytic infiltration and areas of mild perivascular fibrosis were noted. No follicular plugging or pseudocyst formation was found (Figure 2). We prescribed 100 mg/day oral doxycycline and a single intralesional triamcinolone acetonide injection. We revisited the patient at three and eight weeks post-treatment. Our outcome assessment criteria for treatment response were hair regrowth and decreased lesion size/volume. At both follow-ups, there were dramatic and significant improvements considering these criteria (Figure 3).

## CASE DISCUSSION

Alopecic and aseptic nodule of the scalp (AANS) was first reported in Japan in 1992 as a pseudocyst

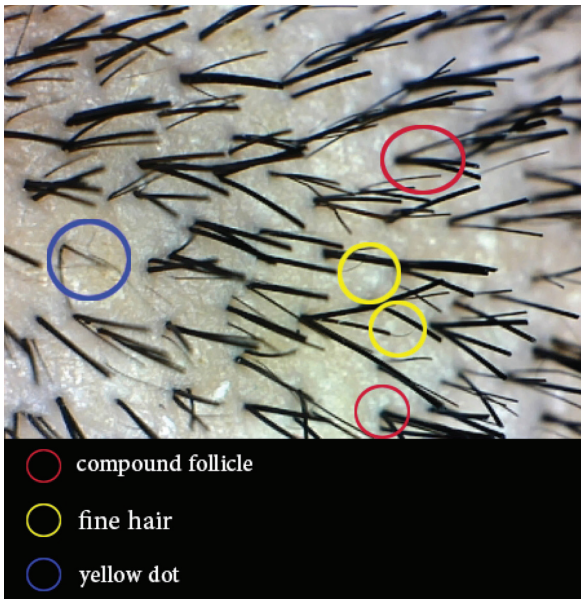


**Figure 1.** Trichoscopy showing compound follicles, yellow dots, and fine vellus hair.

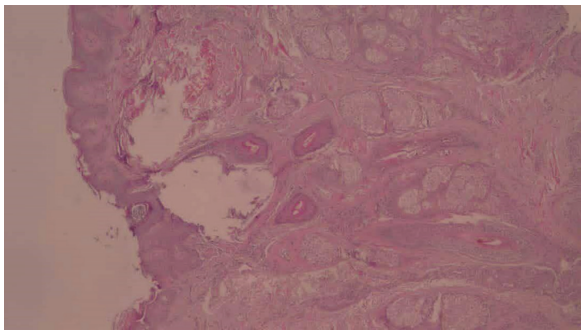

**Figure 2.** Histopathology with H & E staining showing nonspecific changes including an acanthotic, hyperkeratotic epidermis with dermal interstitial and perifollicular lymphohistiocytic infiltration and areas of mild perivascular fibrosis.

of the scalp (PCS). It was named PCS because of cystic cavities with no real cystic walls in histopathologic studies [1]. The first cases in Western populations were reported by Chevallier [2], where the lesions were called "non-infectious abscesses and balding scalp". Tsuruta and colleagues reported four cases with the same features as the cases of Chevallier [3]. In 2009, Abdennader and Reygagne coined the term AANS [4]. In 2011, Abdennader *et al.* reported 15 cases in a prospective study [5], while Seol *et al.* reported a clinicopathologic and therapeutic analysis of 11 Korean cases [6]. Table 1 summarizes the characteristics of the reports found in the literature. Most reported AANS cases were males under 40 years of age. The exact prevalence of AANS is unknown, though most patients have been of Asian or Caucasian ethnicity.

The etiology of AANS is unknown. Some hypotheses consider follicular occlusion as the cause of the nodule (pseudocyst) formation, where this disorder occurs as a particular form of deep folliculitis that leads to nonscarring alopecia [7]. On the other hand, some authors consider a granulomatous reaction secondary to immune response (caused by follicular alteration or a foreign body or some unknown factor) as the cause. The granulomatous infiltrate is probably located around the lower part of the hair follicle, beneath the bulge; this might be the cause of nonscarring alopecia [8].

The majority of reported cases had one or two nodules. Perilesional skin is often normal, and nodules are asymptomatic or can occasionally cause discomfort [9]. The consistency can be firm or fluctuating [6]. Hair loss occurs in an early phase


**Figure 3.** (a) A skin-colored, non-scarring, mobile, alopecic nodule in the right parietal vertex with moderate fluctuation on palpation, surrounded by the normal scalp. (b) Lesion after three weeks. (c) Lesion after eight weeks. (d) Cutis verticis gyrata.

coinciding with the formation of the nodule, healing without scarring. The most common areas are the vertex, followed by occipital and parietal regions, though any scalp area can be affected [9].

The aspirate can be clear serous, yellow, citrine yellow, pinkish, or even purulent depending upon the type of the inflammatory infiltrate and the type of vessels affected by the infiltrate [4,10]. Cultures are always sterile with no bacterial/fungal growth [6].

The histopathologic findings are nonspecific. In one of Abdennader's studies, a granuloma in the deep dermis was found in nearly half

**Table 1.** Characteristics of the reported cases of AANS in the literature

| Study | N | Age, years | M:F | Ethnicity | Number of nodules | Localization | Histopathology | Treatment |
|---|---|---|---|---|---|---|---|---|
| Iwata *et al.*, 1992 | 19 | 18-40 | 14:5 | Asian | 1 | Parietal & frontal | Pseudocyst & granuloma | Aspiration + IL CS (10/19) |
| Chevallier, 1998 | 3 | 17-35 | 2:1 | Caucasian | 1 | N/S | Follicular cyst & granuloma | Repetitive punctures |
| Tsurata *et al.*, 2005 | 4 | 20-29 | 2:2 | Asian | 1 | Parietal & vertex | Pseudocyst & granuloma | Surgical excision |
| Abdennader & Reygagne, 2009 | 18 | 12-38 | 18:0 | Caucasian | ≤2 | Vertex & occipital | Inflammatory infiltrate in reticular & deep dermis + granulomas in deep dermis | Doxycycline 100 mg/d |
| Abdennader *et al.*, 2011 | 15 | 18-49 | 14:1 | Caucasian: 11<br>Black: 4 | Variable ≤ 2<br>6= 1 nodule<br>5= 2 nodules<br>3= 3 nodules<br>1= 6 nodules | Vertex & occipital | Inflammatory infiltrate in reticular & deep dermis, <1/2 with granulomas in deep dermis | Doxycycline 100 mg/d for 3 months |
| Lee *et al.*, 2011 | 1 | 72 | 0:1 | Asian | 1 | Temporal | Pseudocyst-like | Surgical excision |
| Eisenberg, 2012 | 11 | 16-48 | 3:8 | Caucasian: 5<br>Hispanic: 1<br>Arab: 2<br>Black: 2<br>Asian: 1 | 10= 1 nodule<br>2= 2 nodules | Vertex (10) & occipital (1) | N/S | Puncture & IL CS (triamcinolone acetonide 2.5 mg/ml) |
| Fischer-Levancini *et al.*, 2014 | 1 | 16 | 1:0 | Hispanic | 1 | Parietal | Granulomas | Doxycycline 100 mg/d for 8 wks + puncture |
| Gargallo *et al.*, 2015 | 1 | 20 | 1:0 | Hispanic | 2 | Occipital & temporal | Granulomas | Doxycycline 100 mg/d + systemic CS for 2 wks |
| Seol *et al.*, 2016 | 11 | 11-35 | 11:0 | Asian | 10= 1 nodule<br>1= 2 nodules | N/S | Inflammatory infiltrate in deep dermis + granulomas | Doxycycline + IL CS for all |
| Rodriguez-Lobato *et al.*, 2017 | 1 | 11 | 0:1 | Caucasian | 1 | Vertex | N/S | Topical 3% indomethacin solution |
| Fischer-Levancini *et al.*, 2018 | 1 | 34 | 1:0 | Hispanic | 2 | Vertex | Superficial & deep perivascular mixed inflammation & deep chronic folliculitis | Doxycycline 100 mg/d for 8 wks |

Abbreviations: IL, intralesional; CS, corticosteroid; N/S, not stated

of the cases. In the other half, they reported a nonspecific deep lymphohistiocytic infiltrate, which was usually subacute, well-circumscribed, and nonsuppurative. Fibrosis was seldom observed, and the alopecia was non-scarring [5]. We found an acanthotic, hyperkeratotic epidermis with areas of papillomatosis and also an interstitial and perifollicular lymphohistiocytic infiltration, with areas of mild perivascular fibrosis but no follicular plugging. In contrast to the Japanese studies [1,3] and the study by Lee *et al.* [11] (reporting a pseudocyst-like finding) and in line with the Abdennader *et al.* study [5], we found no pseudocyst formation. We believe the differences in racial factors and different hair types or biopsy depths might explain some of these differences.

Garrido-Colmenero *et al.* [12] reported trichoscopic and 15-MHz ultrasonographic findings. In trichoscopy, they found broken hair shafts, black and yellow dots, and fine vellus hair. Rodrıguez-Lobato *et al.* reported a case in a seven-year-old girl with trichoscopic and 18-MHz ultrasonographic findings. They observed black and yellow dots, fine vellus hair, and broken hair shafts [7]. We found compound follicles, yellow dots, and fine vellus hair in trichoscopy; however, we did not observe black dots or dystrophic hair. The ultrasonographic findings in the studies mentioned above were a well-defined, subcutaneous hypoechoic nodule with a few blood vessels on the base and a hypoechoic lesion with increased intralesional flow on Doppler imaging [7,12]. We did not have access to high-

frequency ultrasonography.

The main differential diagnoses for AANS include conditions with follicular occlusion, such as dissecting cellulitis of the scalp or folliculitis decalvans. Alopecia areata, inflamed trichilemmal/other adnexal cysts, bacterial/dermatophytic folliculitis, congenital lesions such as aplasia cutis, triangular alopecia, sebaceous/epidermal naevi, and benign and malignant tumors (particularly metastatic nodules of the scalp) are other disorders that should be considered [4,5,10].

Cutis verticis gyrata (CVG) is a condition associated with excessive soft tissue proliferation, with undulations of the scalp mimicking cerebral sulci and gyri [13]. Its estimated prevalence is 1:100.000 and 0.026:100.000 in males and females, respectively [14]. CVG can be classified into primary and secondary forms, with the primary form itself subcategorized into non-essential and essential types. The primary non-essential form may be associated with mental retardation, cerebral palsy, epilepsy, cataracts, and blindness, while the primary essential form is not related to such abnormalities [14-15]. Secondary CVG is associated with a large list of disorders such as acromegaly, pachydermoperiostosis, psoriasis, chronic pemphigus, myxoedema, intradermal amyloidosis, etc. [15]. As primary CVG is asymptomatic in most cases and has a benign nature, local hygiene supportive measures would often suffice as treatment. If it poses an aesthetic problem, there are surgical treatment options [16]. After the relevant investigations, our patient was categorized as having primary essential CVG. The patient did not receive any treatment for his CVG except localized hygiene practice recommendations.

There are no established treatment protocols for AANS. Several approaches with different follow-up durations have been reported. Table 1 summarizes the treatments described in the literature. Topical (indomethacin 3% solution), intralesional (intralesional corticosteroids), systemic (oral antibiotics like doxycycline; systemic corticosteroids [17]), and surgical (puncture [18], excision) treatments have all been applied. More recent studies have often used an oral antibiotic (doxycycline) ± an adjunct treatment. We observed a dramatic response to oral doxycycline 10 mg/day for eight weeks plus a single dose of intralesional triamcinolone acetonide at the first visit.

## CONCLUSION

We reported the first case of AANS and primary essential CVG occurring simultaneously in a patient. AANS is a rare and probably under-reported cause of non-scarring alopecia with a rapid and dramatic response to non-surgical treatment and a good prognosis. A better understanding of AANS is necessary for proper treatment and avoidance of unnecessary interventions.

**Conflict of Interest:** None declared.

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