A 9-year-old boy presented with a soft subcutaneous nodule with overlying alopecia on the right parietal scalp of 5 months’ duration that had grown in size, became increasingly alopecic, and was complicated by intermittent pain. An excisional biopsy of the nodule revealed deep dermal mixed inflammation with scattered granulomas. No foreign material, definitive cystic spaces, or cyst wall lining was identified. Special stains including periodic acid–Schiff, Fite acid-fast, and Twort Gram were negative for infectious organisms. His postoperative course was uneventful, and no recurrence of the nodule was reported.

WHAT'S YOUR DIAGNOSIS?

a. alopecic and aseptic nodule of the scalp
b. cylindroma
c. dermoid cyst
d. lipoma
e. pilar cyst

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THE DIAGNOSIS:
Alopecic and Aseptic Nodule of the Scalp

Alopecic and aseptic nodule of the scalp (AANS) is an underdiagnosed condition presenting with one or few inflammatory nodules on the scalp with overlying nonscarring alopecia. The nodules can be soft, fluctuant, or firm and are characterized by negative fungal and bacterial stains as well as cultures. Trichoscopic features such as black or yellow dots, fine vellus hairs, and broken hairs have been reported. Dilated follicular openings may be seen and are termed the Eastern pancake sign, as they resemble the bubble cavities formed during the cooking of atayef. The histologic features of AANS often are nonspecific but show a nodular or pseudocystic, lymphohistiocytic to acute inflammatory component centered in the dermis. Granulomatous inflammation or isolated giant cells have been reported within the deep dermis. In our patient, histopathology revealed admixed acute and granulomatous inflammation within the deep dermis (Figure). Treatment of AANS includes oral antibiotics such as doxycycline, intralesional corticosteroids, or excision.

Although the etiology of AANS currently is unclear, a process of follicular plugging or a deep folliculitis sparing the bulge stem cells has been theorized. Young males are disproportionately affected. It is uncertain how much overlap there is, if any, between AANS and pseudocyst of the scalp, the latter of which primarily is reported in the Japanese literature and demonstrates alopecic nodules between the forehead and vertex of the scalp with pseudocystic architecture and granulomatous infiltration on histopathology.

There are several clinical and histologic differences between AANS and other diagnoses in the differential. Dermoid cysts tend to present at birth, with 70% of cases presenting before the age of 6 years, and without overlying skin changes. They represent a benign entrapment of ectoderm along embryonic closure lines during development. Histologic examination typically will show a squamous-lined cyst within the dermis with associated adnexal structures. Cylindromas are benign neoplasms of eccrine sweat glands named after the histologic presentation of cylinder-shaped basaloid cell populations when cross-sectioned. When cylindromas coalesce on the scalp, they form a distinctive morphology sometimes loosely resembling a turban, giving them the previously more common name turban tumors. Cylindromas appear as slow-growing protuberant tumors that are erythematous or flesh colored. Cylindromas are 9 times more common in females. Pilar cysts have a stratified squamous epithelium lining with a palisaded outer layer and are derived from the outer root sheath of hair follicles. Clinically, pilar cysts are smooth mobile cysts that favor skin with a dense concentration of hair follicles. On palpation, pilar cysts are firm due to their keratinous contents and typically are nontender unless inflamed. Lipomas are benign mesenchymal tumors with mature adipocytes that often appear as subcutaneous nodules without overlying skin changes, though they can involve deep fascia. On palpation, lipomas generally are soft, mobile, and nontender.

REFERENCES


