Granulomatous Inflamation in a Case of Preauricular Sinus Mimicking Lupus Vulgaris: Case Report

Bir Preoriküler Sinüs Olgusunda Lupus Vulgarisi Taklit Eden Granülomatöz Yangısal Yanıt

S. Pelin KARTAL DURMAZLAR, MD,[a]
Fatma ESKİOĞLU, MD,[a]
Hüseyin ÜSTÜN, MD[a]

[a]Dermatology Clinic,
Dışkapı Yıldırım Beyazıt Training and Research Hospital and the Pathology Department, Ankara Training Hospital, Ankara

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Yazışma Adresi/*Correspondence:*
S. Pelin KARTAL DURMAZLAR MD
Dermatology Clinic,
Dışkapı Yıldırım Beyazıt Training and Research Hospital, Ankara,
TÜRKİYE/TURKEY
pelin@dr.com

ABSTRACT Preauricular sinus is a minor developmental anomaly. In most cases, the sinus is unilateral and right-sided. Being a common congenital condition, preauricular sinus may remain asymptomatic throughout the entire life span of patients. Only approximately one third of all asymptomatic patients are aware of their malformation. Its clinical importance is due to the accompanying syndromes that may accompany the condition and to the associated complications. Sometimes a yellowish-white discharge consisting of cellular debris and intermittent secondary infections, most commonly with staphylococcal species are observed. Edema, erythema and discharge are common signs of infection. We presented an unusual clinical presentation of preauricular sinus clinically mimicking lupus vulgaris with a foreign body-type granulomatous inflammation on histopathological examination.

Key Words: Abnormalities; lupus vulgaris

ÖZET Preoriküler sinüs minör bir gelişimsel anomalidir. Birçok olguda sinüs, daha çok sağ tarafta olmak üzere tek taraflı görülür. Sık karşılaşılan bir konjenital anomali olarak hastaların tüm yaşamları boyunca semptom vermeden kalabilir. Hastaların yaklaşık sadece üçte biri bu durumdan haberdar olur. Preoriküler sinüsün klinik önemi, beraberinde görülebilen bazı sendromlar ve beraberinde gelişebilecek komplikasyonlardan ileri gelir. Bazen sinüsten hücre artıklarını içeren sarımsı beyaz bir akıntı geldiği gözlenir ve aralıklı olarak daha çok stafilokok suşları ile olmak üzere ikincil enfeksiyonlar görülebilir. Ödem, eritem ve akıntı enfeksiyonun genel bulgularıdır. Burada görünümü ile lupus vulgarisi taklit eden ve yabancı cisim tipi granülomatöz yangısal değişikliklerle karşımıza çıkan bir preoriküler sinüs olgusu sunulmaktadır.

Anahtar Kelimeler: Konjenital defektler; lupus vulgaris

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Preauricular sinus (PAS) is a minor developmental anomaly. The most accepted theory attributes PAS to defective or incomplete fusion of the six auricular hillocks. It is generally symptom-free and is visible in the preauricular region as a small opening.[1] PAS occurs either sporadically or is inherited in an autosomal dominant pattern with about 85% penetrance.[2] Its clinical importance is due to the syndromes that can accompany and to the associated complications.[2,3] We reported an unusual clinical presentation of PAS, which shows foreign body-type granulomatous inflammation histopathologically and mimicks lupus vulgaris clinically.

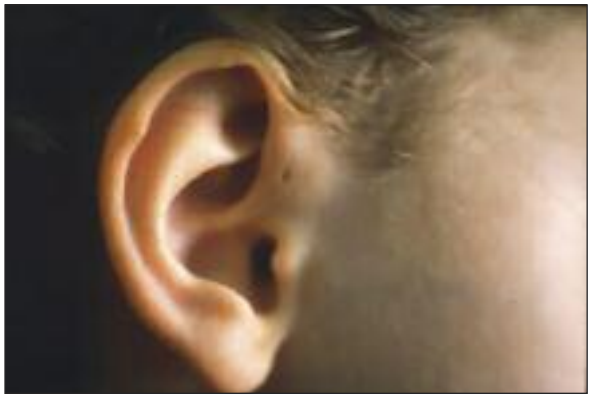

FIGURE 1: Clinical feature of the patient. The auricular pit without signs of inflammation and the preauricular lupus-vulgaris-like plaque. No sinus orifice is discernible within the plaque.

CASE REPORT

A five-year-old female patient was referred to our clinic with a non-healing wound at the anterior side of her right ear of 3-year duration. Her physical examination revealed a well-defined, irregular, red–brown, slightly elevated partially crusted plaque of 1 x 1.5 cm diameter with areas of peripheral atrophy in front of her right ear, between the tragus and the upper anterior helix (Figure 1). We also noticed an auricular pit in the upper anterior helix, apart from the plaque, without signs of inflammation. No sinus orifice was discernible within the plaque. She has had the pit since her birth and pus had started draining from the sinus at the age of one. At the beginning, pus drainage had responded to either topical or systemic antibiotic treatment but by time, a permanent wound had appeared at the anterior side of her ear. Since there were no signs of infection at the site of pit, our first impression of the plaque was that of lupus vulgaris due to its clinical appearance and due to the lack of response to antibiotic therapy and the long duration of the lesion though we could not rule out the possibility of an unusual presentation of PAS. She was a completely healthy child except for this complaint. PAS was also present in her three-year-old brother, her uncle and two of her cousins, but they were asymptomatic (Figure 2). Her complete blood count, sedimentation rate, routine biochemical tests as well as tuberculin test, chest X-ray, cultures of skin lesion, renal ultrasonography, and audiometry were completely normal. After informed consent was obtained from her parents skin biopsy was performed. Histopathological analysis of the punch biopsy specimen demonstrated epithelioid histiocytes, lymphocytes, plasmocytes and foreign body-type multinucleated giant cells (Figure 3). A polymerase chain reaction (PCR) examination revealed no sign of mycobacterial infection. Although sinus tract was not detected on histopathological examination, it was found consistent with symptomatic PAS in which the sinus tract is often replaced by granulamatous inflammatory process.[3] Based on the clinical, histological and laboratory findings, a foreign body-type granulomatous inflammatory process in PAS clinically mimicking lupus vulgaris

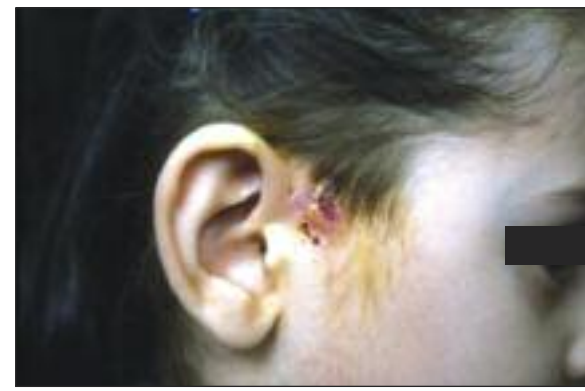

FIGURE 2: Clinical appearance of the patient's 3-year-old brother: Asymptomatic preauricular sinus without signs of inflammation.

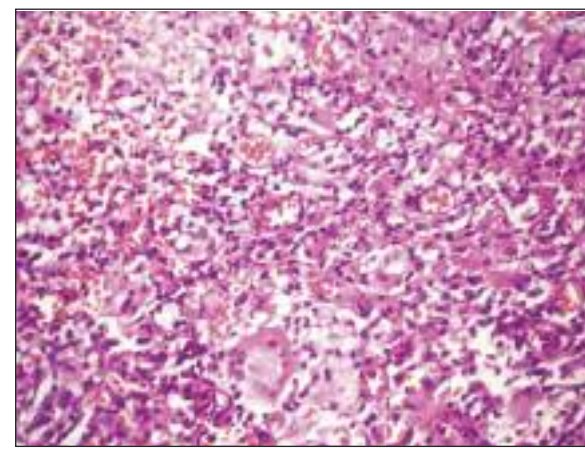

FIGURE 3: Histopathological findings showing epithelioid histiocytes, lymphocytes, plasmacytes and foreign body-type multinucleate giant cells (Hematoxylin-Eosin stain; original magnification x200).

was diagnosed. Surgical excision by an experienced head and neck surgeon was recommended but the family postponed the operation.

DISCUSSION

PAS is a benign congenital malformation of the preauricular soft tissue with an estimated incidence of 0.1-0.9%.[2] Recently, the prevalence of PAS in Taiwanese newborns has been reported to be 0.6%.[4] PAS is usually narrow and short, occasionally arborizing and following a tortuous course near the external ear.[3] A small pit is often noted adjacent to the external ear, usually located at the anterior margin of the ascending limb of the helix.[2,3] The opening of PAS has also been reported along the posterosuperior margin of the helix, the tragus or lobule and postauricular region.[5] PAS and anomalies of the first branchial cleft sometimes share similar clinical presentations. Unlike PAS, anomalies of the first branchial cleft arise from incomplete closure of the ectodermal portion of the first branchial cleft. An opening in the cheek below the angle of the mandible or an opening in the upper neck above hyoid bone suggests an anomaly of the first branchial cleft.[5]

Being a common congenital condition, PAS may remain asymptomatic throughout the entire life span of patients. Only approximately one third of all asymptomatic patients are aware of their malformation.[1] Sometimes a yellowish-white secretion consisting of cellular debris is discharged through the stratified squamous sinus tract to the surface of the skin and intermittent secondary infections, most commonly with staphylococcal species are observed.[1-3] Edema, erythema and pain are common symptoms. In case of a secondary infection, appropriate antibiotics should be administered since the inflammation of the sinus tract leads to its elongation.[1] In most cases, the sinus is unilateral and right-sided.[2] PAS has been described as a part of a number of multiple congenital anomaly syndromes. The most well-known of these is the branchio-oto-renal (BOR) syndrome.[6,7] BOR syndrome consists of conductive, sensorineural or mixed hearing loss, PAS, structural defects of the ear, renal anomalies and failure and lateral cervical fistulas, cysts or sinuses. Other syndromes, some of which have been reported separately but might in fact overlap with BOR, can be associated with PAS as well.[3] In a report, familial occurrence had no association with major congenital disorders, whereas in one third of sporadic cases, serious congenital anomalies could be detected.[8] Leung and Robson carried out a prospective study to investigate the incidence of renal anomalies associated specifically with PAS.[6] They found that, on renal ultrasonography, 3 of 69 children with PAS also had a renal anomaly. They concluded that such anomalies were significantly more common in patients with PAS than the 1% incidence of renal anomalies reported in the general population. They suggested that renal ultrasonography should be performed on all patients with PAS. This opinion is not shared widely though. Another study suggested that a renal ultrasound should be performed in patients with isolated PAS accompanied by one or more of the followings: another malformation or dysmorphic feature, a family history of deafness, auricular and/or renal malformations or a maternal history of gestational diabetes.[9] In the absence of these findings, this study suggested that renal ultrasonography was not indicated. In a study of young adult male subjects with PAS, association with ear and renal abnormalities were rare; only 3 of 10 PAS subjects had renal structural minor abnormalities, which consisted of partial or complete duplication of the pelvicalyceal system.[10] One limitation of this study was that only males were evaluated. In a recent report, it is recommended to seek for associated anomalies in patients with PAS.[2] In the view of literature reports it can be concluded that in selected cases renal ultrasound may be appropriate. Where no associated abnormalities are identified and where PAS is asymptomatic, no further evaluation is indicated. Conduction or mixed type hearing loss is the most common co-existing pathology, so audiometric examination is recommended.[2,3,11]

Many believe that recurrent infection is an indication for performing surgery, while others think that, even asymptomatic sinuses should be excised.[3] Various surgical techniques, aimed at en-

suring complete dissection, have been described in the literature.[2,3,12] The use of methylene blue to demonstrate the sinus tract by probing has been recommended and it reportedly lowers the recurrence rate.[12] Incomplete excision is believed to be the cause of recurrence.[2]

Very rarely chronic inflammation and granulomatous changes may develop at any site of the sinus tract; due to the ruptured sinus wall or chronic irritation of the sinus discharge; which may be in the form of a lupus-vulgaris-like foreign body reaction.[13-15] The importance of such cases is that they can be misdiagnosed. Histologically, stratified squamous epithelium lines the sinus tract: the surrounding tissue can contain plasma cells, lymphocytes and neutrophils. In case of severe inflammation sinus tract is often replaced by granulation tissue.[3]

Our case demonstrates the difficulty in diagnosis, because our first impression was that of a lupus vulgaris plaque due to its clinical appearance, the lack of response to antibiotic medication, and the site and long duration of the lesion. However, mycobacterium culture and PCR examination and latter findings excluded this diagnosis. Besides, the histological examination of our patient revealed granulomatous inflammation, which was consistent with symptomatic PAS in which the sinus tract is often replaced by granulamatous inflammatory process.[3] Therefore, this case is important to remind that chronic sinus infection may lead to unusual clinical manifestations of PAS.

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