

tar pustulosis. 25% of patients, primarily males, experience moderate to severe nodulocystic acne. Other related conditions like HS, Sneddon-Wilkinson, Sweet syndrome and Pyoderma gangrenosum, are less common. Roughly 10% of SAPHO syndrome sufferers experience symptoms like fever and fatigue.⁵ Elevated inflammation markers, leukocytosis and mild anaemia might manifest in lab results. Treatment of SAPHO syndrome is necessary for symptom relief and prevention of further complications such as impairment of bone and joint function. Treatment strategies are often based on evidence from case reports and may vary depending on the disease manifestations in the patient; this is primarily because clinical trials have not been conducted because of its rarity. A nonsteroidal anti-inflammatory drug and short-term glucocorticoids provide initial osteoarticular relief. Methotrexate can treat peripheral arthritis in the absence of axial disease, while TNF inhibitors are preferred for severe enthesitis and axial disease. For patients presenting with both osteoarticular and skin manifestations in SAPHO syndrome, healthcare providers may consider diverse approaches, such as oral retinoids for palmoplantar pustulosis and acne. Viable options for challenging cases include bisphosphonate therapy, Interleukin (IL)-17 inhibition, IL-1 inhibition, IL-12/23 inhibition and Janus kinase inhibition.⁶ Our twin patients' response to adalimumab matches existing literature documenting its efficacy for SAPHO syndrome. This report is unique due to the simultaneous and rare occurrence of AF and HS in monozygotic twins as part of the SAPHO syndrome.

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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Authors' contributions

Ilaria Scandagli: Concepts, design, definition of intellectual content, manuscript preparation.

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Matteo Ruggieri: Definition of intellectual content, literature search, manuscript preparation.

Tommaso Amadori: Manuscript preparation, manuscript editing, manuscript review.



Francesca Prignan: Concepts, definition of intellectual content, manuscript editing, manuscript review, guarantor.

Conflicts of interest

None declared.

References

1. Sallés M, Olivé A, Perez-Andres R, Holgado S, Mateo L, Riera E, et al. The SAPHO syndrome: a clinical and imaging study. *Clin Rheumatol.* 2011;30:245–9.
2. Naik HB, Cowen EW. Autoinflammatory pustular neutrophilic diseases. *Dermatol Clin.* 2013;31:405–25.
3. Hayem G. SAPHO syndrome. *Rev Prat.* 2004;54:1635–6.
4. Depasquale R, Kumar N, Lalam RK, Tins BJ, Tyrrell PNM, Singh J, et al. SAPHO: what radiologists should know. *Clin Radiol.* 2012;67:195–206.
5. Nguyen MT, Borchers A, Selmi C, Naguwa SM, Cheema G, Gershwin ME. The SAPHO syndrome. *Semin Arthritis Rheum.* 2012;42:254–65.
6. Firinu D, Murgia G, Lorrain MM, Barca MP, Peralta MM, Manconi PE, et al. Biological treatments for SAPHO syndrome: an update. *Inflamm Allergy Drug Targets.* 2014;13:199–205.

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Pigmented mammary Paget disease: a diagnostic challenge[☆]



Dear Editor,

Mammary Paget disease is a rare type of adenocarcinoma. It is closely associated with breast cancer.¹ Pigmented mam-

mary Paget Disease (PMPD) is an uncommon variant of Paget disease that can mimic melanoma clinically, dermoscopically, and histopathologically due to the presence of melanin pigment.² Here, we present a case of PMPD with dermoscopic melanoma and Bowen's Disease (BD)-like findings, in which histopathological examination made a clear distinction.

A 45-year-old female was admitted to our outpatient clinic with a slowly expanding, asymptomatic, erythematous skin lesion with some pigmented areas on the left nipple that had been present for one year. Upon dermatological

[☆] Study conducted at the Kayseri City Education and Research Hospital, Kayseri, Turkey.

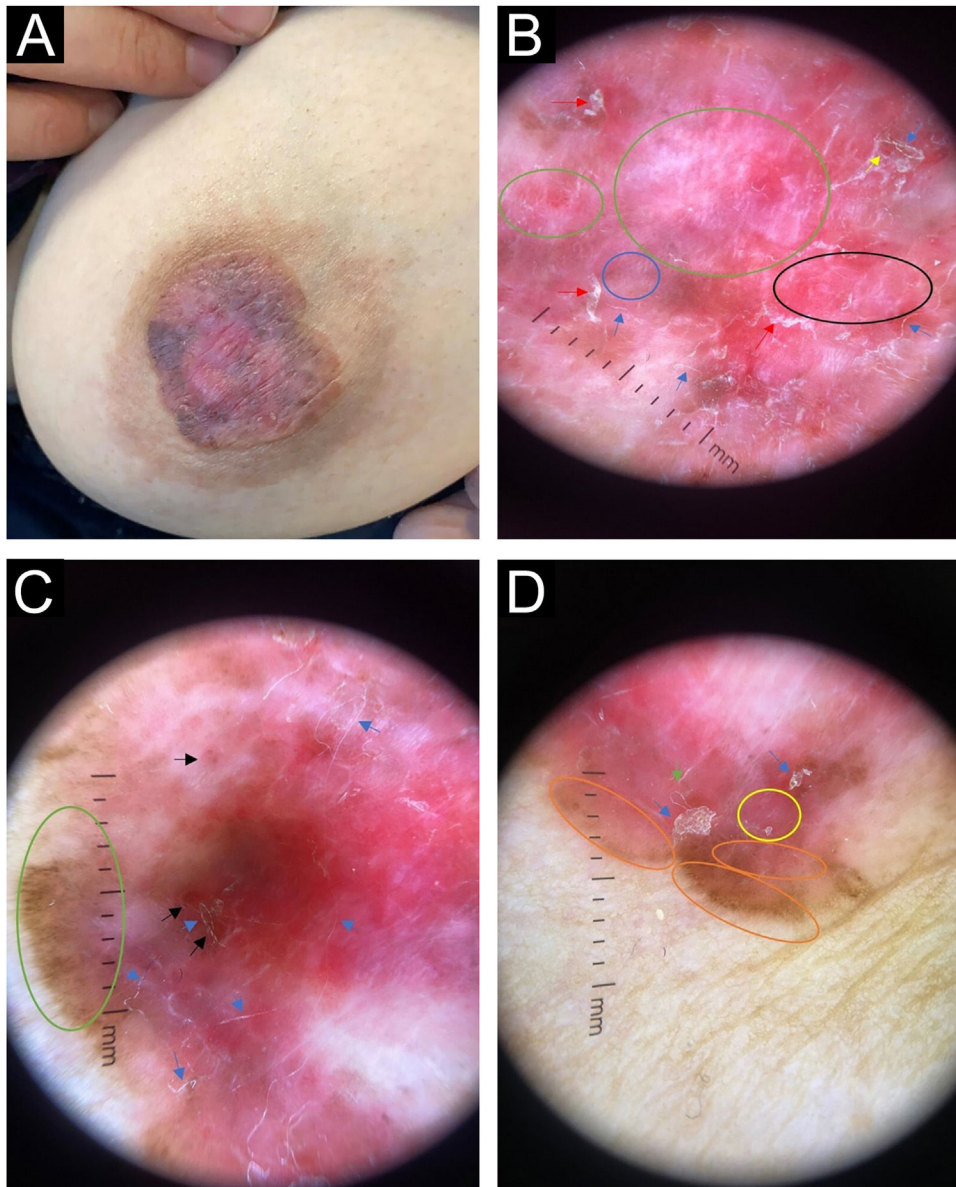


Figure 1 (A) Brown-pink plaque on the left areola measuring 3 × 3 cm in size, with irregular borders, and flattened nipple. (B) Red arrow: scale, blue arrow: adherent fabric fibers, yellow arrow: superficial erosions, green ring: shiny white streaks, blue ring: irregular dotted vessel, black ring: pink, white structureless area. Dermlite DL4- ×10 magnification - polarized mode. (C) Blue arrow: adherent fabric fibers, black arrow: superficial erosions, orange arrow: scale, green ring: segmental brown radial lines and brown-gray dots. Dermlite DL4- ×10 magnification - polarized mode. (D) Blue arrow: scale, green arrow: adherent fabric fibers, yellow ring: dotted vessels, orange ring: segmental radial lines and brown-gray dots. Dermlite DL4- ×10 magnification - polarized mode.

examination, a 3 × 3 cm brown-pink plaque was found on the left areola, and the nipple was flattened. The plaque exhibited centrifugal growth, asymmetry, irregular borders, and scattered scaling (Fig. 1A). Dermoscopy revealed a chaotic lesion, with numerous clues for melanoma, including a pink, white structureless area, central white lines, and segmental brown radial lines. There were also brown-gray dots arranged in lines and dotted vessels in lines in some areas, adherent fabric fibers, and superficial erosions which are characteristic of Bowen's disease (Fig. 1B–D). Although the clinical and dermoscopic findings were inconclusive, they were suggestive of pigmented Paget disease, melanoma,

and pigmented BD. The histopathological examination of the areolar incisional biopsy revealed a glandular epithelium with large pale cytoplasm, large nuclei, and prominent nucleoli that were arranged both singly and in clusters in the epidermis. Immunohistochemically, tumor cells were pan Cytokeratin (CK) and CK 7 positive, Estrogen Receptor (ER), and mucin negative (Fig. 2A–C). Histopathology was consistent with Paget's disease based on these findings. The nipple skin biopsy of the patient, who underwent breast-conserving surgery by the general surgeon, was compatible with Paget's disease, and underlying high-grade ductal carcinoma in situ was detected.

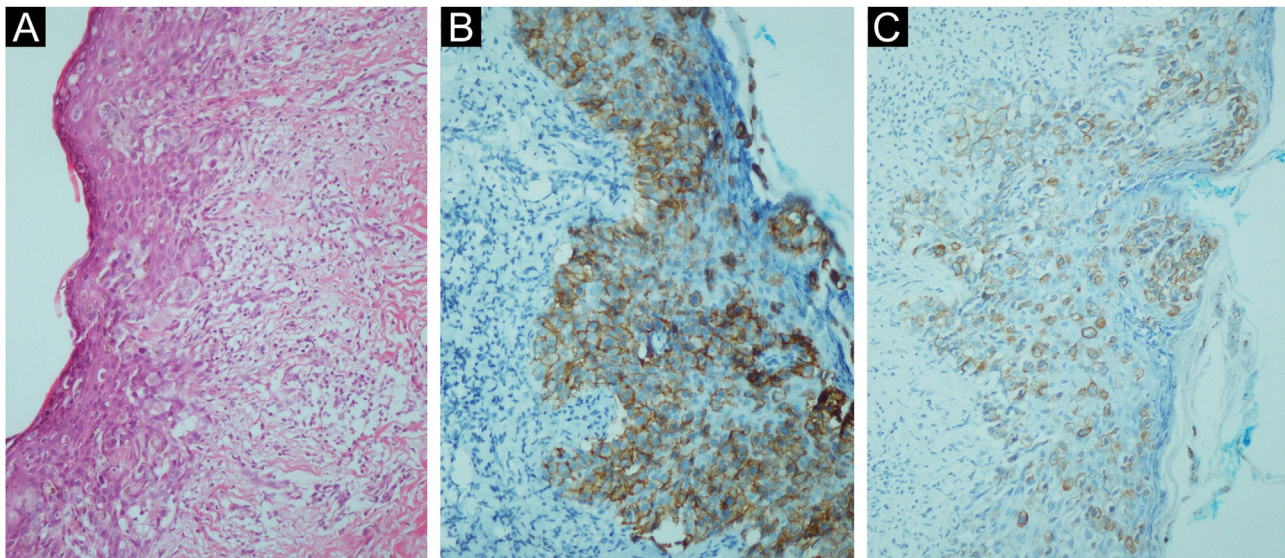


Figure 2 (A) Malignant epithelial cells dispersed as single or clustered cells in the epidermis (Hematoxylin & eosin, $\times 200$). (B) Membrane staining of malignant epithelial cells in the epidermis with C-ERB-B2 ($\times 200$). (C) Cytoplasmic staining of malignant epithelial cells with CK7 ($\times 200$).

Paget disease of the nipple is a rare type of intraepidermal adenocarcinoma. It accounts for only 1%–4% of all breast malignancies.^{1,3} Dermoscopically, the most common findings of non-pigmented mammary Paget disease include white scales, pink structureless areas, dotted vessels, erosion/ulceration, and white shiny lines. On the other hand, the typical findings of PMPD are gray granules and dots, pink structureless areas, and white lines.⁴

The differential diagnosis of PMPD encompasses a large spectrum of conditions from melanoma to BD and lentigo/melanosis of the nipple and areola.⁴ Segmental brown radial lines, pink-brown structureless areas, scaling, clustered or linearly arranged brown dots, and dotted and coiled vessels can also be observed in pigmented BD. BD of the nipple is an extremely rare condition that resembles nipple melanoma; however, the dermoscopic characteristics have not been fully described in the literature.^{4,5} In the medical literature, fewer than 20 cases of nipple melanoma have been published. Clinical, dermoscopic, and even confocal microscopy findings cannot reliably distinguish PMPD from melanoma.⁴

In our case, dermoscopic features are shared by both melanoma and BD, dermoscopic differentiation was not possible. In conclusion, this case once again demonstrated how difficult it can be to make a dermoscopic diagnosis of PMPD and that melanoma, Paget disease, and BD can manifest similar clinical findings or similar macroscopic findings. Therefore, in suspected cases, a comprehensive approach that includes histopathological and immunohistochemical examinations, in addition to dermoscopy, is necessary for an accurate diagnosis.

Authors' contributions

Esratur Ünal: The study concept and design, writing of the manuscript, critical review of the literature (The physician

who examines the patient, takes the biopsy, takes dermoscopic photographs, writing of the manuscript).

Bengü Nisa Akay: Critical review of important intellectual content, final approval of the final version of the manuscript (The physician who gives advice in writing the case and evaluates dermoscopic findings).

Gökçen Gündoğan: Data collection, analysis and interpretation (Performing histopathological examination).

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References

1. Crignis GSN, Abreu LD, Buçard AM, Barcaui CB. Polarized dermoscopy of mammary Paget disease. *An Bras Dermatol*. 2013;88:290–2.
2. Akay BN, Demirdag HG, Tasolar MK, Serel S, Kirmizi A. Pigmented mammary Paget disease mimicking cutaneous melanoma. *Int J Dermatol*. 2018;57:e49–52.
3. Bhat YJ, Bashir S, Wani R, Hassan I. Dermoscopy of paget's disease. *Indian Dermatol Online J*. 2020;11:674–5.
4. Apalla Z, Errichetti E, Kyrgidis A, Stolz W, Puig S, Malvehy J, et al. Dermoscopic features of mammary Paget's disease: a retrospective case-control study by the International Dermoscopy Society. *J Eur Acad Dermatol Venereol*. 2019;33:1892–8.
5. Cameron A, Rosendahl C, Tschandl P, Riedl E, Kittler H. Dermatoscopy of pigmented Bowen's disease. *J Am Acad Dermatol*. 2010;62:597–604.

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Plaque psoriasis on the tongue: case report[☆]



Dear Editor,

Although psoriasis is a disease with considerable prevalence in the Brazilian population, isolated oral involvement is rare. The histopathology of oral psoriasis was initially described in 1903 by Oppenheim,¹ and over the years a few more reports have improved the literature on this subject.²⁻⁴ The diversity of clinical presentations and the occasional isolated occurrence, without association with skin lesions, are factors that complicate the diagnosis.^{2,3} The present report describes a case of tongue psoriasis without associated skin involvement.

A previously healthy 45-year-old woman, a dentist, complained of a whitish plaque on her tongue for three months, with progressive increase. She showed no response to the use of triamcinolone acetonide orabase ointment for short periods or to the use of nystatin. She had a geographic tongue before the plaque appeared. She was an alcoholic and denied any family or previous history of psoriasis, continuous use of medication, or smoking. Physical examination revealed whitish plaques on the sides of the tongue (Fig. 1); there was no evidence of skin or skin appendage

lesions. Histopathology (Fig. 2) showed hyperplastic squamous mucosal epithelium with elongation of the epithelial ridges, parakeratosis, and exocytosis of neutrophils with formation of Munro's microabscesses. The lamina propria exhibited a predominantly lymphocytic inflammatory infiltrate, congested vessels, and edema. No fungi were identified and therefore the diagnosis was consistent with tongue psoriasis. Treatment involved interruption of alcohol consumption and betamethasone elixir three times a day and there was good progress after six months of treatment (Fig. 3).

Psoriasis is a multifactorial and chronic disease, the etiology of which has yet to be completely elucidated. The most common oral mucosa findings are fissured tongue and geographic tongue, occasionally as an isolated manifestation.²⁻⁴ Other possible clinical presentations are yellowish-white or circinate plaques with histopathology compatible with psoriasis.^{2,3} Oral psoriasis is frequently mistaken for other more common diseases, such as lichen planus, candidiasis, and syphilis, which makes the diagnosis challenging.² In cases of isolated oral lesions, clinical suspicion must always be confirmed by anatomopathological examination.

Histopathology is similar to that of cutaneous psoriasis, with findings secondary to the hyperproliferation of keratinocytes: hyperkeratosis, parakeratosis and hypogran-



Figure 1 Whitish plaques on the tongue.

[☆] Study conducted at the Hospital das Clínicas da Universidade Federal de Minas Gerais, Belo Horizonte, MG, Brazil.