Mongolian spots combined with halo-like disappearance surrounding café au lait spots

Sir,

Phacomatosis pigmentovascularis is a rare group of disorders characterized by the presence of a capillary malformation and a pigmented nevus. It is a poorly understood sporadic disorder explained by somatic mutation triggered by mosaicism or a result of twin spotting by one or more somatic mutation causing dysregulation in multiple embryonic cell lines, including melanocytes and fetal vasculature. Mongolian spots with superimposed café au lait spots and halo-like disappearance surrounding each spot have been reported in only a small number of patients. Here, we describe a new case of this rare phenomenon.

A 5-month-old baby was seen for evaluation of multiple skin lesions on her body at the Faculty of Medicine, Akdeniz University, Antalya, Turkey. She was born at term (birth weight 3 kg, height 50 cm) after a normal pregnancy to unrelated parents. At birth, she had a homogenous grayish-blue patch covering the lumboSacral region and buttocks. It was consistent with Mongolian spot. After 1 month, multiple brownish macules appeared and gradually increased in size and number on the extremities and on the buttocks within the Mongolian spot. Dermatologic examination revealed a halo nevus-like appearance of normal skin color around café au lait macules on the Mongolian spot [Figure 1a and b]. On clinical examination, she was healthy and had reached all the developmental milestones appropriate for her age with no abnormalities on physical and neurological examination. She had no signs of neurofibromatosis such as axillary freckling, peripheral or central nervous systems anomalies, poliosis circumscripta, kyphoscoliosis, bone hypertrophy or pseudoarthrosis. Neurofibromatosis type-I FISH test was analyzed for Neurofibromatosis-1 and showed no deletion in the 17q11.2 region. Family history was negative for signs or symptoms of neurofibromatosis-1, and clinical examination of the parents was normal.

Three punch biopsy specimens were obtained from three areas: the blue patch, the brown macule and the halo-like zone surrounding this macule [Figure 2a-c]. In halo-like zone and brown macule (café au lait macule), dermal melanocytes were small, round and their degree of melanization was less than that in the blue patch (Mongolian spot). Further, dendrites of dermal melanocytes were decreased in the halo-like zone and in the dermis of the café au lait macule [Figure 3a-c]. All three specimens had no evidence of inflammatory reaction.

This phenomenon has been reported rarely in the literature. Although two of the patients had neurofibromatosis and one had phacomatosis cesioflamma, others had no sign for genetic disorders. Our case did not show any other sign for any systemic disorder or genetic defect for neurofibromatosis.

Mongolian spots can be seen in association with nevus flammeus (phacomatosis pigmentovascularis type II) or with cutis marmorata telangiectatica congenita (phacomatosis pigmentovascularis type V). Wolf et al., differently from previously reported phacomatosis cases, described an unusual case with Mongolian spots and segmental café au lait macules. They proposed the name “phacomatosis pigmento-pigmentalis” in analogy to other similarly named entities such as phacomatosis pigmentovascularis and phacomatosis pigmentokeratotica. We also think that Mongolian spots combined with café au lait macules was not a simple coincidence, but the result of twin-spotting phenomenon. The reason for the halo phenomenon is unknown. This halo-like zone has normal skin color and does not show any evidence of inflammatory reaction in biopsy specimens, unlike the depigmentation in halo nevus. The most specific histologic finding of all cases is that dendrites of dermal melanocytes were not clearly seen in the halo-like zone and the dermis of the café au lait macule. Dendrites of dermal melanocytes of Mongolian spots seemed to disappear at the site of café au lait macule and in the halo-like zone.

In conclusion, phacomatosis pigmentovascularis is a group of disorders with different clinical features. Halo-like disappearance of surrounding café au lait spots on Mongolian...
Multiple halo nevus-like zone of normal skin color around café au lait macules on the Mongolian spot.

The dermal pigmented melanocytic proliferation was slightly more prominent in the blue spot biopsy [hematoxylin and eosin (H and E), ×400].

Dermal melanocytes were small, round, and their degree of melanization was less than in the Mongolian spot (H and E, ×400).

Diffuse pan-dermal dendritic and pigmented melanocytosis was more prominent in the blue patch biopsy (MelanA, ×400).

No inflammatory infiltrate was noticed in the biopsy from the halo-like zone (H and E, ×400).

spot is a very rare feature of this group. There is no specific treatment for this phenomenon at present. After reviewing similar cases in the literature in detail, we suggest the term “phacomatosis pigmento-halo-pigmentalis” to better name this
unusual condition. Better understanding of this phenomenon and the process involved can help us arrange a follow-up program to diagnose its accompanying systemic or dermatologic involvement and treat these lesions in the future. Therefore, documentation of more such cases is essential to confirm this suggested entity.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

Aslı Bilgiç Temel, Cumhur Ibrahim Bassorgun¹, Banu Nur², Erkan Alpsoy³

Department of Dermatology and Venereology, Besehir State Hospital, Konya, Departments of ¹Pathology, ²Pediatric Genetics and ³Dermatology and Venereology, Faculty of Medicine, Akdeniz University, Antalya, Turkey

Correspondence: Dr. Aslı Bilgiç Temel,
Department of Dermatology and Venereology,
Besehir State Hospital, Konya, Turkey.
E-mail: asli.bilgictemel@saglik.gov.tr

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that these tumor cells were diffusely and strongly positive. Immunohistochemical investigations revealed the destruction of appendages in the entire dermis (both superficial and deep dermis) with infiltration by round-to-oval cells in diffuse sheets involving Sweet's syndrome, nodular amyloidosis or leprosy. A skin metastatic thyroid nodules, cutaneous lymphoma, sarcoidosis, the clinical differential diagnoses considered were cutaneous presentation showed a total white cell count of 48,000 with 14% blasts. In the peripheral smear then. However, peripheral blood counts done at 12,000/mm³ on the fourth postoperative day, which was considered pre-surgically, on the first postoperative day and reduced to 12,000/mm³ on the fourth postoperative day, which was considered normal except for a total leukocyte count of 10,200/mm³ within normal limits at the time of the surgery. His blood count was undergone a surgical procedure of total thyroidectomy and anterolateral neck dissection for invasive papillary carcinoma of the thyroid. The patient had no family history of any malignancy. The patient could recall the presence of a solitary swelling over his forehead even before undergoing a surgical procedure of total thyroidectomy and anterolateral neck dissection for invasive papillary carcinoma of the thyroid – A rare case of cutaneous myeloid sarcoma in a patient with invasive papillary carcinoma of thyroid – A rare case of cutaneous myeloid sarcoma in a patient with invasive papillary carcinoma of thyroid. Coexistent cutaneous myeloid sarcoma in a patient with invasive papillary carcinoma of thyroid – A rare case of cutaneous myeloid sarcoma in a patient with invasive papillary carcinoma of thyroid. Studies of cutaneous involvement in myeloid myelodysplastic syndromes, acute myeloid leukemia, myeloproliferative neoplasms and myeloid leukemia. Myeloid sarcoma is reported in 2.5% to 9.1% of patients with acute myeloid leukemia. As per the World Health Organization, myeloid sarcoma is a tumor occurring at a site other than bone marrow. Also known as granulocytic sarcoma or chloroma (as it produces a greenish color due to the presence of enzyme myeloperoxidase), it is an extramedullary mass consisting of myeloid blasts with or without maturation, as the central nervous system, lymph nodes, gastrointestinal tract, liver, lungs, spleen, oral mucosa and testes. They were hard in consistency, immobile, nontender and organomegaly. His hepatic, renal and metabolic parameters were all normal. The patient had no family history of any malignancy. The patient could recall the presence of a solitary swelling over his forehead even before undergoing a surgical procedure of total thyroidectomy and anterolateral neck dissection for invasive papillary carcinoma of the thyroid. The presence of a solitary swelling over his forehead even before undergoing a surgical procedure of total thyroidectomy and anterolateral neck dissection for invasive papillary carcinoma of the thyroid indicates acute transformation of the underlying bone marrow disease and carries a poor prognosis of cutaneous myeloid sarcoma. Other sites reported to be involved as acute myeloid leukemia, myeloproliferative neoplasms and chromosomal abnormalities, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

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