Correspondence

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There are less than 10 case reports in literature.

Another hallmark of this case was the presence of the carcinoma at the esophageal-gastric junction which to the best of our knowledge was not reported elsewhere. Another very important highlight of our case was the severe dysphagia seen in our case. Moderate to severe dysphagia is the hallmark of DM due to the involvement of the cricopharyngeus and other skeletal muscles of the pharynx and esophagus. Hence, this symptom is considered to be a part and parcel of DM and usually OGD-scopy is not done. However, in our case, the severe symptoms and dysphagia to both solid and liquid foods prompted us to do OGD-scopy and we could pick up the esophageal carcinoma. Hence, we advocate that an OGD-scopy be made mandatory for any patient with DM to rule out esophageal carcinoma.

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Conflicts of interest

There are no conflicts of interest.

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Figure 4: Biopsy of the esophagus showing positive stain for cytokeratin indicating a primary carcinoma of the esophagus (cytokeratin stain, ×100)

Figure 3: Biopsy of the esophagus showing cells with hyperchromatic nuclei, scanty cytoplasm, pleomorphic cells, and bizarre cells (black arrows) suggestive of poorly differentiated carcinoma, (H and E, ×400)

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Neurofibromatosis Type 1 with Becker’s Nevus and Nevus of Ota

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Sir,
The co-occurrence of neurofibromatosis type 1 (NF-1), Becker’s nevus (BN), and nevus of Ota is expected to be very rare. No such cases were found in MEDLINE and EMBASE.
A 22-year-old woman, who had a 5-year history of underlying slowly growing painless soft masses involving her trunk, presented to us with an asymptomatic congenital brownish patch on her right cheek [Figure 1a and b]. There had been multiple café-au-lait macules (CALMs) of different size all over the body [Figure 1c]) since her birth and bilateral axillary freckles appeared when she was 5 year old. The soft skin-colored nodules which were found initially during adolescence increased both in number and size gradually, and also the brownish patch of her right face became heterogeneous and presented excess hair partly. There was no history of musculoskeletal or neurological disorder. Systematic examinations (skeletal survey, ophthalmological and ENT evaluation, etc.) were done and no other defect was found. Both her mother and maternal grandfather suffered from NF-1.

Histological examination of one of the nodules on her abdomen revealed a tumor consisting of wavy cells arranged in a haphazard manner in fibrous and myxoid stroma [Figure 2] which indicated neurofibroma. So she was diagnosed as NF-1 associated with BN and nevus of Ota. We recommended Q-switched laser treatments, but she refused.

NF-1, caused by mutation of the gene neurofibromin 1 (located at chromosome 17q11.2), is a multisystem disorder affecting around one in 3,500 individuals. Various kinds of hyperpigmentation skin disorders are related to NF-1, including CALMs, hyperpigmentation overlying plexiform neurofibromas and axillary freckles, nevus spilus, congenital giant melanocytic nevus, and segmental unilateral melanosis, etc.[1,2]

BN is a unilateral, hyperpigmented, hairy cutaneous disorder which is usually associated with various pigmented lesions, malignant melanoma, and leiomyoma too. It is characterized by lesions located over the upper trunk and displays a corresponding regional relationship to the nevus.[3] Many cutaneous complications, including lymphangioma, intradermal or connective tissue nevi, and perforating folliculitis have been reported in association with this disorder.[3]

Nevus of Ota is a congenital or acquired pigmentary disease of the skin and mucous membranes, both of which are innervated by the trigeminal nerve. It is rare that NF-1, BN, and nevus of Ota occurred simultaneously[1,2,4] and the pathogenesis of hyperpigmentation related to NF-1 is still unclear.

There had been intimate relationships between the above three disorders from the perspective of developmental biology. In fact, NF-1 is a multisystem disorder caused by abnormal development of neural crest cells from which melanocytes and Schwann cell precursors (SCPs) are derived. In addition to contributing primarily to glia, SCPs adjacent to nerves were a cellular source of melanocytes and postnatal mature pigmented melanocytes in the cutis.[5] SCPs losing contact with the peripheral nerves during development acquired a melanoblast fate.[6] Consequently, BN and nevus of Ota might just be different manifestations of hamartoma arising from the differentiation of neural crest cells in NF-1. More studies are needed to focus on the coexistence of lesions especially in terms of malignancy in the future.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and anonymity cannot be guaranteed.

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There are no conflicts of interest.
Unusual Periungual Nodulocystic Lesions Leading to the Diagnosis of Extranodal Marginal Zone Lymphoma

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Sir,

We report a 91-year-old female who was examined for a 6 month lasting periungual nodulocystic lesions with dystrophic nails affecting several digits of the upper extremities [Figure 1a]. Lesions were painful. Patient was on oral antidiabetics and antihypertensive medication, otherwise she was healthy. She had been treated by another dermatologist with systemic itraconazole with the suspicion of candidal paronychia with no effect. General blood count was normal. Biochemistry examination revealed slightly elevated uric acid. We consulted a rheumatologist who excluded gout and periungual lesions as tophi. General physical examinations including chest X-ray and abdomen ultrasound were normal. We performed a punch biopsy with the result of extranodal marginal zone B-cell lymphoma [Figure 2a and b]. The patient had no systemic symptoms of lymphoma. She was immediately examined by a hematologist. Lymphoma cells were found in microscopy of peripheral count examination by the hematologist and we concluded that the skin lesions were secondary cutaneous manifestations of systemic B-lymphoma. Because of the age of patient, no further tests were performed to examine if any other organs were affected (gastric mucosa, colon, etc). The serology for Helicobacter pylori and Borrelia burgdorferi were negative. The patient declined a bone-marrow examination. The systemic therapy with repeated cycles [Figure 1: (a) Nodulocystic periungual lesions of the digits of the upper extremities. (b) Regression of the lesions after systemic therapy]

Histopathological findings. Diffuse dermal infiltrate composed of small CD20+ B lymphocytes. (a) (H and E, x40); (b) (Anti CD20 immunohistochemistry, x200)

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