diagnosed as EBA by skin biopsy, direct and indirect immunofluorescence, and enzyme-linked immunosorbent assay. The patient was treated with rituximab at a weekly dose of 375mg/m² for 4 weeks. Treatment was well tolerated without any side effects during rituximab therapy. Complete remission was achieved and sustained during 6 months follow-up period. So far, seven cases of EBA treated successfully with rituximab have been reported. From these observations, rituximab can be considered a treatment option in patients with recalcitrant and severe EBA and more clinical studies will be needed to assess the efficacy and adequate dose of rituximab in the treatment of EBA.

키워드: Epidermolysis bullosa acquisita, Rituximab

A mild form of mucopolysaccharidosis type 2 (Hunter syndrome) with characteristic skin lesions

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A 6-year-old male child presented with multiple symmetric skin colored papules and nodules that coalesce into a reticular pattern over the scapula, lateral aspect of the upper arms and thighs. At first, the lesions appeared 2 years ago and had gradually increased in number over time, but had not triggered symptoms such as itching or pain. In addition, persistent Mongolian spot on the buttock, mild hepatomegaly, claw-hand deformities and puffy eyelids were also noted. The punch biopsy specimen obtained from a typical nodule of the right upper arm revealed separated collagen bundles in the dermis. No specific abnormalities were noted in the epidermis and appendage. Alcian blue staining showed pale bluish material between collagen fibers, consistent with mucin. An enzyme assay performed on blood leukocyte showed deficiency of iduronate-2-sulfatase, confirming the diagnosis. As results of these findings, the patient was diagnosed with Mucopolysaccharidosis type 2, Hunter syndrome.

키워드: Hunter syndrome, Mucopolysaccharidosis type 2

Fibro-osseous pseudotumor of the digit mimicking verruca vulgaris

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Fibro-osseous pseudotumor of the digit (FOPT) is a benign, subcutaneous fibroblastic proliferation with foci of osseous differentiation, usually developing on the fingers and toes of young to middle-aged adults. Herein, we report a case of fibro-osseous pseudotumor of the digit which resembled verruca vulgaris. A 32-year-old man presented with a 6-month history of a nodular lesion with eroded surface on his right second toe. The clinical impression at the initial visit was verruca vulgaris and two sessions of cryotherapy was attempted. However, it revealed underneath a bone-like protrusion circumscribed by hyperkeratotic skin. Skin biopsy was performed, and it demonstrated well-formed mature bony trabeculae of cortical type with minor portion of woven bones, rimmed by active osteoblasts in the background of fibrocollagenous proliferation. These findings were consistent with FOPT and the lesion was treated by local excision without recurrence.

키워드: Fibro-osseous pseudotumor of the digit, Verruca vulgaris, Fibroblastic proliferation

Propylthiouracil induced DRESS

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DRESS((Drug Rash with Eosinophilia and Systemic Symptoms) is a hypersensitivity drug reaction which is characterized by cutaneous eruption and hematologic abnormalities and systemic involvement. A 49-year-old woman had presented with generalized erythematous confluent maculopatches for 4 days. She had a facial edema and mild fever with cervical lymphadenopathy. But we could not find any signs of infection. Laboratory tests showed a eosinophilic leukocytosis and elevated level of liver function tests. On her past history, she was diagnosed with Graves’ disease and treated with
propylthiouracil since 1 month ago. A skin biopsy specimen showed necrotic keratinocytes and lymphocytic infiltration in the upper dermis and perivascular area. She discontinued medications and was treated with systemic steroid. After stopping the medication, her skin lesions were gradually subsided and liver function and hematologic abnormalities were improved, also. Based on these findings, we diagnosed this case as a PTU induced DRESS. Clinicians should aware DRESS not only because of its often delayed diagnosis, but also its life-threatening potential. Herein, we report propylthiouracil as a drug may induce a DRESS, but has never been reported yet in Korea.

키워드: Propylthiouracil, DRESS

Malignant fibrous histiocytoma occurred during interferon α-2b therapy for malignant melanoma

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Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma of middle and late adulthood. The most common sites of origin are the proximal extremities, particularly the thigh and buttock. About two-thirds of the tumors occur in striated muscle and less than 10% are confined to the subcutis. A 60-year-old women underwent right index finger amputation for stage IIC malignant melanoma a year ago. Since then she has received continuous treatment with interferon α-2b and there was no evidence of recurrence. Two months ago, she noted a hard growing tender nodule on her right forearm. Excisional biopsy was performed and histopathologic examination was consistent with MFH. Recently, rare case report of dermal origin was reported, and it is more difficult to find a report of cutaneous malignant fibrous histiocytoma occurred during interferon α-2b therapy for malignant melanoma. In this case, we can assumed that interferon therapy may have influenced the occurrence of MFH or genetic predisposition like DNA repair system defect may predispose to her two different cutaneous malignancies. The exact reason for occurrence of MFH in this patient is obscure and needs to be examined further.

키워드: Malignant fibrous histiocytoma, Interferon, Melanoma

Superficial angiomyxoma

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Superficial angiomyxoma is a clinically variable, infrequent, benign myxoid tumor of soft tissue. They are clinically characterized as slow-growing, asymptomatic nodules or polyps located on the trunk, lower extremities, and head and neck of adults. Their histopathological features include poorly delimited multinodular tumors containing spindled and stellate fibroblasts admixed with thin-walled blood vessels and embedded in a mucinous stroma. A 25-year-old male presented with a 6-year-history of a protruding mass on the left thigh. The lesion was a single skin-colored, hard, non-movable mass protruding on the posterior aspect of the left thigh. The patient complained of no symptoms and the tenderness was not noted. An ablative laser surgery was performed once 4 years before presentation. He had no family history of similar skin lesions. The excisional biopsy specimen explained a myxoid tumor containing spindled and stellate cells with vessels in the dermis. The immunohistochemical study revealed negative stain for SMA, desmin, S-100, Ki-67 and CD68 and positive stain for CD34. The patient was diagnosed with superficial angiomyxoma. We present this interesting case with a brief literature review.

키워드: Superficial angiomyxoma

Two cases of nicolau syndrome in treated with Non-steroidal anti-inflammatory drug injection therapy

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Nicolau syndrome, also known as livedoid dermatitis or embolia cutis medicamentosa, is a rare cutaneous adverse drug reaction characterized by the acute onset of cutaneous and soft-tissue necrosis following intramuscular drug injection. The typical presentation is pain around the injection, followed by erythema, livedoid patch, finally necrosis of skin, subcutaneous