

cases have also been reported. It has been the source of considerable controversy since their original description because of marked differences in definitions from previous reports. So, this syndrome may be the best to characterize by a wide spectrum of clinical phenotypes from familial atypical multiple mole-melanoma syndrome to 10 or fewer or even a single DN without a personal/family history of melanoma. Its importance is that it identifies an at-risk population group for the subsequent development of melanoma. Herein, we report 4 Korean patients who fulfilled clinical and histopathological characteristics of DNS (3 with sporadic and 1 with familial). All had several to multiple, clinically-atypical and histopathologically-confirmed DNS scattered on the whole body. Melanoma developed in two patients. In Korea, with extended criteria of DNS, several underreported cases of DNS can be found since 1988. So more cases of DNS are expected to be found in Korea with thorough examination.

키워드 : Atypical mole, Dysplastic nevus, Dysplastic nevus syndrome, Korean

FC I-13

Intralesional 3% sodium tetradecyl sulfate for treatment of cutaneous Kaposi's sarcoma

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Kaposi's sarcoma is an angioproliferative disease thought to be from endothelial cell lineage, classically described as a multipigmented sarcoma appearing on the lower extremities of elderly men. Radiation therapy is commonly used, and other treatment modalities include systemic chemotherapy and surgical excision. However, these treatments can induce severe complications, impairing the immune system of patients. In comparison, sodium tetradecyl sulfate (STS) cause endothelial surface damage, which in turn induces an inflammatory reaction, which leads to sclerotization of vessels. It also cause less complication than other systemic treatments, and is low in cost. A 96-year-old woman presented with hard, violaceous indurated plaques and protruded erosive papules on right dorsum of foot and ankle, diagnosed as Kaposi's sarcoma by biopsy. Due to her old age, local treatment instead of chemotherapy or aggressive operation was started. Cryotherapy was tried on plaque lesions, and intralesional injection of 3% STS on nodular lesions, respectively. 3% STS

treated lesions changed color from purple to black immediately, but no associated pain or other unwanted effects were reported. Five sessions of treatment were done and the lesions showed definite shrinkage without any complications for 3 months. Our case indicates that this sclerosing agent is an effective alternative drug for the treatment of nodular Kaposi's sarcoma lesions.

키워드 : Kaposi's sarcoma, Sodium tetradecyl sulfate, Cryotherapy

FC I-14

Metastatic malignant melanoma presenting as agminated spitz nevus

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A 36-year-old male patient presented with multiple erythematous waxy papules and nodules on his right medial thigh. He had a history of amputation of his right second toe due to malignant melanoma (stage IIa) 3 years ago. At the time of the surgery of primary tumor, right inguinal lymph node dissection revealed no nodal involvement. 3 years after the diagnosis of primary tumor, crops of multiple erythematous papules and nodules newly developed. Histopathologic evaluation of the papule showed compound nevus consisting of nests of epithelioid cells in wedge shaped arrangement, reminiscent of Spitz nevus. However, cytologic features, including high mitotic figure, variable cellularity, and some hyperchromatic nuclei, raised concerns about melanoma. In addition to the pathologic findings, the tumors were on the right thigh, the same side as the primary malignant melanoma. The patient underwent wide excision of the tumor with split thickness skin graft.

키워드 : Cutaneous metastatic malignant melanoma, Spitzoid melanoma

FC I-15

Two cases of clear cell sarcoma with different clinical and genetic features

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Clear cell sarcoma, also known as malignant melanoma of soft parts, is a rare malignancy with melanocytic differentiation. The tumor is usually found in the lower extremities of young adults with being deep seated and is characterized by multiple local recurrences with late metastases. Sometimes, it is difficult to diagnose clear cell sarcoma only with histological studies. In that case, cytogenetic analysis can be helpful. Recurrent t(12;22)(q13;q12), resulting in a EWS-ATF1 fusion, is identified in most case of clear cell sarcoma and helps distinction clear cell sarcoma from metastatic melanoma or other spindle cell tumors. But the other molecular characters are not well known yet. Herein, we report two patients with clear cell sarcoma with different clinical and genetic features. The first one's tumor was located in dermis and the other one's was located in subcutaneous fat layer. Interestingly, we found BRAF mutation in dermal type and KIT mutation in subcutaneous type. BRAF and KIT is well known mutations for inducing oncogenesis of cutaneous melanoma. But, on the contrary, these mutations are known to be not common in clear cell sarcoma. With these cases, we suggest the gene mutations in tumorigenic process also can be a key factor in determine the characters of clear cell sarcoma, too.

키워드 : BRAF, Clear cell sarcoma, KIT, Melanoma

FC I-16

A novel mutation (c.3717del5) in COL7A1 in a patient with recessive dystrophic epidermolysis bullosa

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Dystrophic epidermolysis bullosa (DEB) is a rare hereditary bullous disease characterized by sublamina densa tissue separation and abnormalities in the anchoring fibrils, resulting from COL7A1 mutations. The clinical features of DEB vary broadly in severity, from relatively mild, localized blistering of the extremities to generalized blistering, mutilation, and esophageal blistering. To date, over 550 mutations have been identified within COL7A1 in different variants of the DEB group. Here we present a patient with recessive DEB caused

by compound heterozygous mutations of a p.Q1211X (c.3631C>T) mutation in one allele, and a novel deletion mutation (c.3717del5) in the other allele of COL7A1. A 17-day-old female patient presented with bullae and erosions on the whole body. She had difficulty to thrive and showed multiple milia formation and loss of toenails. Histological examination revealed subepidermal blister and labeling of dermal-epidermal junction with anti-type IV collagen antibodies revealed linear basement membrane zone fluorescence on the roof of separated skin. Electron microscopy showed only some fibrillar structures with a wisp-like morphology below the lamina densa. Mutation analysis of the genomic DNA extracted from the patient and her parents identified two mutations in the type VII collagen gene: the novel c.3717del5 mutation and the c.3631C>T mutation, each of them also found in the parents separately.

키워드 : Dystrophic epidermolysis bullosa, Novel mutation

FC I-17

Subepidermal calcified nodule on sole

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Calcinosis cutis is classified into 4 categories: dystrophic, metastatic, iatrogenic, and idiopathic. Idiopathic type is calcification with no apparent tissue damage or demonstrable metabolic effect. Subepidermal calcified nodule is an unusual type of idiopathic calcinosis cutis and usually presents as an asymptomatic, solitary, firm, verrucous, white or yellowish nodule. It can commonly occur in young children with previous trauma history. It is generally seen on the face and neck especially eyelid, cheek, but sole is very rare site. Four patients present as several-months-history of solitary 0.2~0.3mm sized firm nodule on the sole. The age of most patients was below 1 year old and there were no previous history of trauma except 1 case. Biopsy specimens were obtained from the soles and the histopathologic examination revealed hyperkeratosis, acanthosis and amorphous basophilic material in cystic space beneath the epidermis that means deposition of calcium salt. These histopathological features were consistent with subepidermal calcified nodule. Serum calcium and phosphorus levels of the patients were within normal limits. Herein, we present 4 cases of subepidermal calcified nodule on sole that is atypical location.

키워드 : Sole, Subepidermal calcified nodule