Figure 1. Ill-demarcated, irregularly-pigmented macular on the proximal nail fold before imiquimod therapy (A), 6 weeks (B), and 18 weeks into treatment (C). (D) Dermoscopic image of the lesion. Atypical pigment network with bleeding. (E) A skin biopsy showed full thickness dysplasia of the squamous epithelium with hyperkeratosis and parakeratosis stained with HE. (F) After imiquimod therapy, the tumor cells were not observed. Original magnification x200 (E, F), high magnification view (x400) is shown on right lower panel (E).

well as precancerous lesions including Bowenoid papulosis and BD [4, 5]. Moreover, the HPV detectable rate is high in digital BD [6]. Although no type of HPV-DNA was detected in our patient, including HPV16, 18 and HPV-60, which is detected in pigmented warts, we considered the BD lesion was resolved by imiquimod treatment, based on its antitumor effects by activating innate and acquired immunity.

In summary, topical imiquimod 5% cream is an effective alternative treatment option for BD patients who are unsuitable for other treatments like surgery. Further studies to figure out an optimal dosing scheme are required.


Vascular-type Ehlers-Danlos syndrome presenting as recurrent compartment syndrome

The vascular type of Ehlers-Danlos syndrome (EDS type IV, EDS-IV) is an autosomal dominant disorder caused by structural defects in the procollagen type III collagen encoded by the COL3A1 gene [1]. EDS-IV is clinically characterized by thin translucent skin, extensive bruising and a distinctive facial appearance, including prominent eyes, small lips, a pinched nose, and hollow cheeks. It may result in premature death because of arterial rupture, gastrointestinal perforation or rupture, or uterine rupture [2]. Acute compartment syndrome of the extremities by arterial rupture has rarely been described in EDS-IV [3]. Herein, we report a case of EDS-IV presenting as recurrent compartment syndrome caused by rupture of an aneurysm.

A 33-year-old Japanese man presented as an emergency to our Department of Orthopedic Surgery with the sudden onset of painful swelling of his left forearm. His grandfather had suffered from a cerebral hemorrhage. His parents were in good health. He had experienced a spontaneous pneumothorax at age 18 and had developed compartment syndrome of the left lower leg, probably due to the rupture of an aneurysm two years previously. Magnetic resonance imaging revealed rupture of the aneurysm of the left ulnar artery and surrounding massive hematoma. The fascia was incised, and the aneurysm was resected (figure 1A). Examination revealed prominent eyes, pinched nose...
The glycine 183 serine substitution in type III procollagen. From these findings, the mutation is novel, but this may not affect the triple-helix structure of procollagen. The Pro337Ser mutation would prevent the formation of a triple-helix structure of type III procollagen. From these findings, the diagnosis of EDS-IV was confirmed [1]. We did not analyze the genes of his parents. The glycine 183 serine substitution in type III procollagen has been reported in at least seven families of EDS-IV [2], but their clinical features were unspecified. Although no significant correlation between the nature or the location of the mutation and the type or the frequency of the major complication has been reported [2, 4, 5], further accumulation of EDS-IV cases and data regarding COL3A1 gene mutation may give new insight into the genotype-phenotype correlation. Arterial rupture is a major complication of EDS-IV and may lead to death. The mean age at onset of this arterial complication is 25 years [2]. About half of the EDS-IV cases involved thoracic or abdominal arteries, and the rest were divided equally between those in the head and neck and those in the extremities [2]. Arteries in the lower extremities, especially the femoral arteries, were more frequently involved than those in the upper extremities [4]. This vascular complication can result in enlargement of the aneurysm, hematoma, and compartment syndrome. Compartment syndrome is a condition characterized by increased pressure within a closed space of the body. Once a diagnosis of compartment syndrome is made, urgent surgery is necessary to prevent muscle and nerve damage. A major cause of compartment syndrome of the extremities is trauma, including bone fracture and deep burn, whereas rupture of an aneurysm is a rare cause [3]. If a patient with compartment syndrome is already known to have a diagnosis of EDS-IV, quick examination and vascular surgery are possible. Routine medical examination and prophylactic measures, including the avoidance of contact sports or isometric sports, would decrease the number of life-threatening vascular complications.

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**Figure 1.** A) Fasciectomy and removal of hematoma of the left forearm showed ruptured aneurysm of the left ulnar artery. B) Facial appearance including prominent eyes and a pinched nose. C) Hyperextensibility of the left thumb.