

well-established. It is known that SpSCCs that are associated with radiation, burn scars, and immunosuppression may correlate with aggressive clinical courses<sup>5</sup>. Further studies of this rare entity are needed to establish its biological behavior, and the accumulation of more case reports will aid in determining a precise prognosis.

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# Bilateral Segmental Neurofibromatosis on the Face

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Dear Editor:

Neurofibromatosis is a heterogeneous disorder clinically characterized by the presence of neurofibromas, multiple café-au-lait spots, intertriginous freckles, and Lisch nodules<sup>1</sup>.

A 62-year-old woman presented with diffusely scattered papules on the forehead that had been present for 10 years. She had no family history of neurofibromatosis. On physical examination, asymptomatic, soft, flesh-colored papules of 3~5 mm diameter were observed to be distributed over the ophthalmic branch of the right and left trigeminal nerves (Fig. 1A, B). There were no other abnormalities in any other body region. Her general physical examination revealed a normal status, including intelligence, speech, auditory function, and visual acuity.

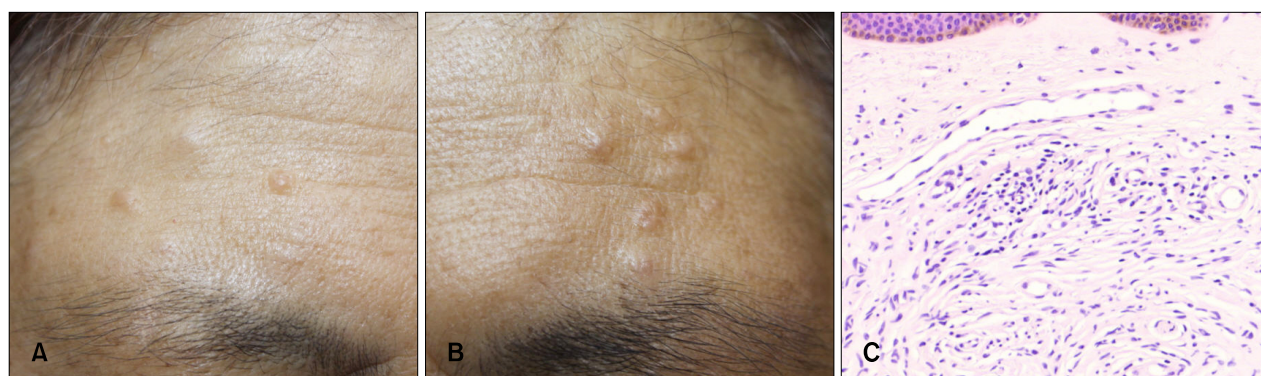
Histopathological examination showed a well-circumscribed tumor in the dermis with a normal overlying epidermis. The tumor consisted of loosely spaced spindle-shaped cells and wavy collagenous strands in the myxoid stroma. Nuclear pleomorphism and mitoses were not observed (Fig. 1C). Overall, the features were consistent with a neurofibroma. The papules on the forehead were excised for cosmetic reason.

Segmental neurofibromatosis is a rare form of neurofibromatosis that is characterized by café-au-lait macules and neurofibromas, or only neurofibromas, distributed in only one dermatome, and less commonly in two or more dermatomes<sup>1</sup>. Segmental neurofibromatosis was categorized into four subtypes by Roth et al. in 1987: a true segmental form (type I), a localized form with deep involve-

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**Fig. 1.** (A, B) Asymptomatic, multiple, soft, skin-colored papules on the right and left forehead. (C) Spindle-shaped cells and eosinophilic, thin wavy collagenous strands in the myxoid stroma (H&E,  $\times 200$ ).

**Table 1.** Review of cases of segmental neurofibromatosis involving the face reported in the English-language literature

Case	Sex/age (y)	Age of onset (y)	Café-au-lait spots	Location	Dermatome	Classification by Roth et al.	Reference no.
1	Male/45	Childhood	—	Right cheek, nose, and mucosa of the lip and nostril	V 2	Type I	1
2	Female/51	Not described	—	Left ear, cheek, nose, chin, and upper neck	V 2, 3	Type I	1
3	Female/35	Childhood	—	Right lower chin and anterior neck	V 3	Type I	1
4	Male/22	6	—	Left side of the nose and adjacent parts of the cheek	V 2	Type I	5
5	Female/59	19	+	Left periorbital region, upper and lower lids, cheek	V 1, 2	Type I	3
6	Male/38	33	—	Left cheek	V 3	Type I	4
7	Male/61	39	—	Central forehead, both cheeks, nose, nasolabial area	V 1, 2	Type III, IV	5
8	Male/37	Childhood	—	Central forehead, nasolabial area	V 1, 2	Type III, IV	5
9	Female/12	Childhood	—	Nasolabial area	V 2	Type III, IV	5
10	Female/62	52	—	Right forehead, left forehead	V 1	Type IV	Our case

ment (type II), a hereditary segmental form (type III), and a bilateral segmental form (type IV). Our case belongs to the bilateral segmental type. Dermatologists should initially perform a physical examination to investigate for cutaneous manifestations elsewhere on the body and an ophthalmologic examination to detect Lisch nodules, in order to determine the presence of either generalized disease or segmental neurofibromatosis. No specific management guidelines exist for segmental neurofibromatosis; however, patients with the segmental type need to be assured that they do not have the generalized form of neurofibromatosis-1 and that they have a low risk of developing any disease-related complications<sup>2</sup>. Hager et al.<sup>1</sup> reviewed 82 cases of segmental neurofibromatosis. The neurofibromas were mostly unilateral; however, bilateral neurofibromas were reported in five patients. The affected der-

matomes were the cervical (31 patients), thoracic (33 patients), lumbar (20 patients), and sacral (5 patients) dermatomes. To date, only nine cases of segmental neurofibromatosis on the face have been reported in the English literature<sup>3-5</sup> (Table 1). To our best knowledge, this is probably the first reported case in the Korean literature of segmental neurofibromatosis involving the face. We report a case of bilateral segmental neurofibromatosis with an unusual location.

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## Retiform Purpura Caused by the Use of Cocaine, That Was Probably Adulterated with Levamisole

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Dear Editor:

Retiform purpura is a dermatological condition characterized by reticulated, stellate, or serpentine shaped purple lesions on the skin and mucous membranes. New, multiple cases of retiform purpura after the use of levamisole adulterated cocaine have been reported. Levamisole is an anthelmintic drug with immunomodulatory and immunostimulating properties. It has been used in humans to treat rheumatoid arthritis, cancer of the colon and nephritic syndrome in children. It was withdrawn from use in the United States in 2000 because of the risk of agranulocytosis<sup>1</sup>.

We report the case of a 52-year-old woman receiving treatment with levothyroxine for hypothyroidism. Two days after consuming cocaine, she developed painful skin lesions with arthralgia on both wrists. Physical examination revealed plaques and papules infiltrated to touch, purpuric on the edges and necrotic in the center, with reticular and stellate lesions on both cheeks, the tip of the

nose, outer left ear, and lower limbs (Fig. 1). Biopsy revealed thrombotic vasculopathy of the small and medium blood vessels in the dermis and subcutaneous cell tissue (Fig. 2). Blood tests revealed leukopenia, neutropenia, and lymphopenia. Antinuclear antibodies (ANA, titer 1 : 1.280) in anti-neutrophil cytoplasm antibodies (ANCAs) against myeloperoxidase with a p-ANCA pattern (titer, >100 [0 ~ 5]), and ANCAs against proteinase 3 with a c-ANCA pattern (titer, 6.8 [0 ~ 5]) were also found. Hypocomplementemia of C3 was detected. The tests for thrombosis and coagulation, serology, cryoglobulins and antiphospholipid antibodies were normal or negative. Cocaine was detected in the urine sample. The results of chest radiography and urine sediment test were normal. A diagnosis of retiform purpura resulting from the use of cocaine, that was probably adulterated with levamisole was made. She was prescribed with low dose oral prednisone. The hematological symptoms cleared 5 days later, after one month, the skin lesions had healed without sequelae.

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