Phakomatosis Pigmentovascularis Associated with Pyogenic Granuloma as well as Sturge-Weber Syndrome and Klippel-Trenaunay Syndrome

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A 21-year-old Korean male was referred to our department in June, 1999 for the evaluation of extensive reddish patches and gray-bluish pigmentation since birth. Physical examination revealed two kinds of patches over the various parts of the body. Reddish patches suggesting nevus flammeus were located on the left side of face, chest, and both upper and lower extremities. Gray-bluish pigmented patches suggesting nevus of Ota were found on both the periorbital areas. A 0.5 × 0.6cm erythematous papule was found on the right anterior chest. The results of its histopathologic examination were compatible with pyogenic granuloma. We made the diagnosis of phakomatosis pigmentovascularis associated with pyogenic granuloma as well as Sturge-Weber syndrome and Klippel-Trenaunay syndrome.
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Key Words : Phakomatosis pigmentovascularis, Pyogenic granuloma

Phakomatosis pigmentovascularis(PPV) is characterized by the distinctive association of an extensive nevus flammeus with a pigmented nevus and, in some cases, of a nevus anemicus[1]. Although PPV is frequently associated with Sturge-Weber syndrome and Klippel-Trenaunay syndrome, to the our best knowledge, the association between PPV and pyogenic granuloma was very rarely made. In fact, only one case was reported by Hagiwara et al. in 1998[2]. We report herein a case of PPV type IIb associated with Sturge-Weber syndrome, Klippel-Trenaunay syndrome, and pyogenic granuloma.

CASE REPORT

A 21-year-old Korean male was referred to our department in June, 1999 for the evaluation of extensive reddish patches and gray-bluish pigmentation since birth. He was born as the second son with a full-term, normal delivery. There had been no abnormalities in the previous pregnancy. The family history of the patient revealed nothing significant. In 1985, at the age of seven, erythematous papule developed on the right anterior chest. In 1986, generalized tonic-clonic seizures occurred first. Since then, seizure disorders continued to date in intervals of 2 to 3 years.

Physical examination revealed two kinds of patches over the various parts of the body(Fig. 1). Reddish patches suggesting nevus flammeus were located on the left side of face, chest, and both upper and lower extremities. Gray-bluish pigmented patches suggesting nevus of Ota were found on both the periorbital areas. Both sclerae were also pigmented. A 0.5 × 0.6cm erythematous papule was found on the right anterior chest(Fig. 2). The right leg measured 4cm longer in length and 6 cm greater in diameter than the left leg. The patient showed mild mental retardation ; According to the KWIS test, full scale IQ was 59.

Laboratory studies, including complete blood cell count, liver and renal function tests were nor-

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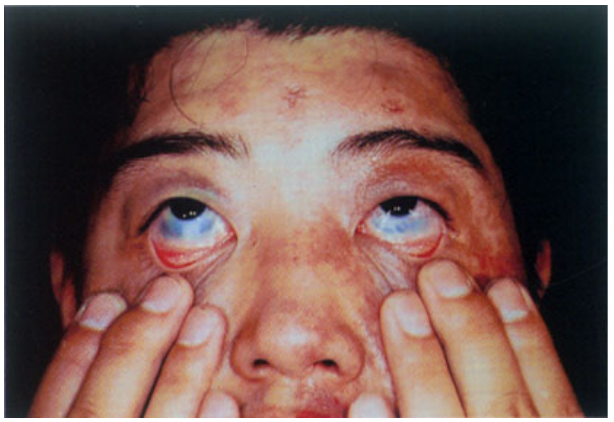

Fig. 1. Two kinds of patches on face : reddish patches on the left side of face and gray-bluish pigmented patches on both periorbital areas and sclerae.

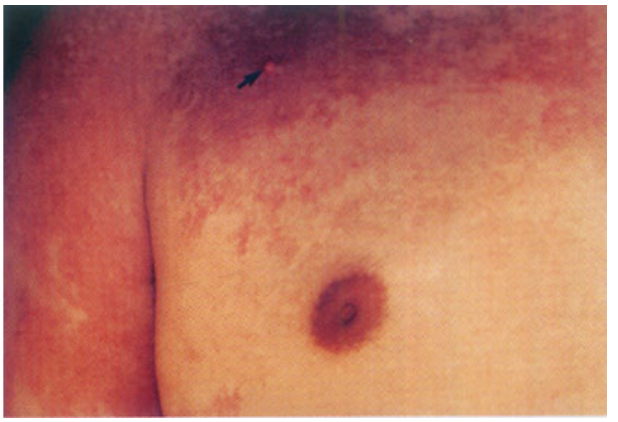

Fig. 2. A 0.5×0.6 cm, erythematous papule was found on the right anterior chest.

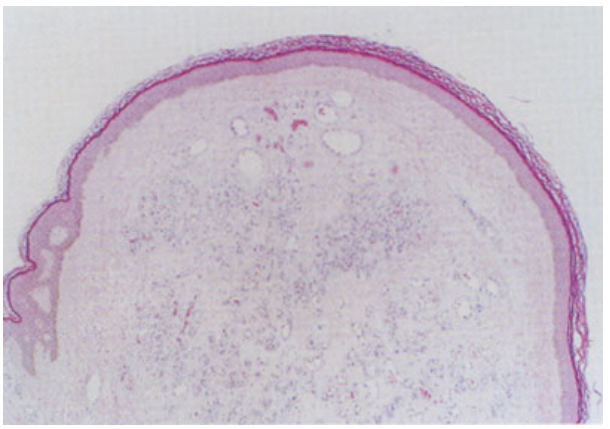

Fig. 3. A biopsy specimen taken from red papule on the right anterior chest showed proliferation of endothelial cells, lobular capillary proliferations, and edema of the stroma(H&E, ×40).

mal. An electroencephalogram demonstrated slow activity of continuously rhythmic delta waves in the left hemisphere. Magnetic resonance imaging of the brain revealed atrophy of the left cerebral hemisphere. He was referred to the ophthalmology clinic of our hospital, where the diagnosis of glaucoma was made. The results of histopathologic examination of reddish patches in the left anterior chest and gray-bluish patches in the periorbital area were compatible with nevus flammeus and nevus of Ota, respectively. We made the diagnosis of PPV associated with Sturge-Weber syndrome and Klippel-Trenaunay syndrome.

A biopsy specimen of an erythematous papule in the right anterior chest showed proliferation of endothelial cells, lobular capillary proliferations, and edema of the stroma. The histopathologic findings were compatible with pyogenic granuloma(Fig. 3).

DISCUSSION

In 1947, PPV was first described as a distinctive association of cutaneous hemangioma and pigmentary nevus by Ota et al[1]. Various models of classification of the entity have been proposed[3]. Among them, the most frequently used has been the one presented by Hasegawa and Yasuhara in 1979[4]. According to them, the entity is classified into four types. Each type is further subdivided[5]: (a) when only cutaneous involvement is present and (b) when cutaneous and systemic diseases are found. Nevus anemicus can be additional features in types II, III, IV. In this case, nevus flammeus and blue spots were found concurrently. Of systemic manifestations, Sturge-Weber syndrome and Klippel-Trenaunay syndrome were found. With respect to the pathogenesis of PPV, the most commonly accepted opinion is that it results from developmental abnormalities of the neural crest-derived vasomotor nerves and melanocytes. Abnormalities in the neural regulations of blood vessels may be important in the development of the vascular component of PPV. This may explain the frequent coexistence of nevus flammeus and nevus anemicus. Anomaly in the migration of the neural crest-derived melanocytes would result in nevus of Ota,

nevus spilus, or mongolian spot[6].

Clinical manifestations associated with PPV include Sturge-Weber syndrome, Klippel-Trenaunay syndrome, microtia, chronic thyroiditis, deafness, scoliosis, abdominal lymphangioma, mental retardation, aplastic anemia, chylothorax, IDDM, and very rarely, pyogenic granuloma.

Pyogenic granuloma can be categorized as a disorder of angiogenesis of unknown etiology. Its pathogenesis involves a neovascular response to an angiogenic stimulus, promoting predominant growth of pericytes with less effect on endothelial cells. The fact that it frequently occurs within a nevus flammeus and has a predilection for the head and neck suggests that the unknown angiogenic stimulus is most likely triggered in dense or dilated vascular beds[7]. In spite of this fact, the association between PPV and pyogenic granuloma was very rarely reported because of the rarity of PPV.

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