Glaucoma in Phakomatosis Pigmentovascularis

Chaiwat Teekhasaenee, MD,1 Robert Ritch, MD2

**Background:** Sturge-Weber syndrome, Klippel-Trenaunay-Weber syndrome, and oculodermal melanocytosis are neural crest disorders in which glaucoma is known to occur. Phakomatosis pigmentovascularis is a neural crest disorder that is found almost exclusively in Asians and has not been described previously in the ophthalmic literature.

**Methods:** The authors describe nine patients with combined oculodermal vascular malformations (five pigmentovascularis, two Klippel-Trenaunay-Weber, two Sturge-Weber) and oculodermal melanocytosis.

**Results:** Ocular melanocytosis was present bilaterally in seven patients and unilaterally in two. Of the 16 hyperpigmented eyes, 13 also had episcleral vascular malformations (EVM). Congenital glaucoma developed in all 10 eyes that had total melanocytosis and EVM. Ocular hypertension developed in one eye with diffuse melanocytosis but partial EVM in childhood. Glaucoma did not develop in one eye with ocular melanocytosis but not EVM.

**Conclusion:** When oculodermal melanocytosis and nevus flammeus (phakomatosis pigmentovascularis) occur together, with each extensively involving the globe, there is a strong predisposition for congenital glaucoma. When one or both are present with only partial involvement, elevated intracocular pressure may develop later in life, and patients should be followed-up at regular intervals for the development of glaucoma. The vascular malformations appear to play a more important role in the predisposition to glaucoma than does the oculodermal melanocytosis. *Ophthalmology* 1997;104:150-157

Oculodermal melanocytosis (ODM, nevus of Ota) is characterized by hyperpigmentation of the eye or facial skin or both in the distribution of the ophthalmic, maxillary, and, occasionally, the mandibular divisions of the trigeminal nerve. When hyperpigmentation of the globe is present in the absence of dermal hyperpigmentation, the term *ocular melanocytosis* (OM, melanosis oculi) has been used.

Sturge-Weber (SW) syndrome (encephalotrigeminal angiomatosis) is a congenital vascular malformation (nevus flammeus or port-wine stain) involving the facial skin, usually in the distribution of the trigeminal nerve, and the cerebral leptomeninges.1

Klippel-Trenaunay-Weber (KTW) syndrome is a congenital disorder consisting of a triad of capillary vascular malformation (nevus flammeus), varicose veins and/or arteriovenous fistulas, and soft tissue and/or bone hypertrophy.2-4 Patients with KTW syndrome often have an associated SW syndrome.5,6

Phakomatosis pigmentovascularis (PPV) is another condition with combined extensive cutaneous vascular malformations (nevus flammeus) and pigmentary nevi (persistent aberrant Mongolian spots or nevus spilus).7 Four types have been described.7-9 They are subgrouped further into cutaneous or systemic disease.10 This disorder is found almost exclusively in Asians and has not been described previously in the ophthalmologic literature.

Congenital glaucoma may occur in infants with ODM,11,12 KTW,5,12-14 or SW15-19 syndrome. Isolated case reports of patients with combinations of these syndromes have appeared in the nonophthalmologic literature, but ocular findings were not reported or were sketchy in most cases. Table 1 is a summation of all of the cases in the
Table 1. Reported Cases of Phakomatosis Pigmentovascularis, Klippel Trenaunay Weber Syndrome, or Sturge-Weber Syndrome with Oculodermal Melanocytosis

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Sex</th>
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<th>FM</th>
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<td>R</td>
<td>–</td>
<td>R</td>
<td>–</td>
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<td>OOU</td>
<td>OOD</td>
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<td>R L and L</td>
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<td>R and L</td>
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</table>

BVM = body vascular malformation; BM = body melanocytosis; FVM = facial vascular malformation; FM = facial melanocytosis; EVM = episcleral vascular malformation; OM = ocular melanocytosis; GL = glaucoma; PPV = phakomatosis pigmentovascularis; NA = not available; † = mentioned the presence or absence; KTW = Klippel Trenaunay Weber Syndrome; SW = Sturge-Weber Syndrome; R = right; L = left; OD = right eye; OOM = oculodermal melanocytosis; OS = left eye; OU = both eyes.

* Diagnosis made retrospectively after reanalysis of cases.  
† Left facial involvement of mandibular distribution only.

By reanalysis of the previously published cases, Glaucoma in patients with PPV has been mentioned in passing, but neither clinical details were noted nor an etiologic connection made. In this article, we describe the detailed ocular findings in nine patients with PPV.

Patients and Methods

Between December 1986 and August 1995, 281 consecutive Thai patients with ODM underwent complete ocular examination on the Glaucoma Service of Ramathibodi Hospital, Bangkok. Nine were found to have phakomatosis pigmentovascularis. Physical examination and computerized tomography of the brain were performed. Medical and family histories were recorded. The patients have been followed-up regularly for the development of glaucoma. Patients with uncontrolled glaucoma underwent goniotomy or trabeculectomy.

Results

The findings in the nine patients are summarized in Tables 2 and 3. Five of the nine patients (cases 1–3, 7, 8) had PPV with extensive cutaneous vascular malformations and pigmented spots involving the face and body. These were termed PPV-diffuse for purposes of analysis. Four patients (cases 4–6, 9) had KTW syndrome with the additional finding of ocular or oculodermal melanocytosis. These were termed PPV-localized.

Congenital glaucoma developed in all nine eyes (cases 1–6, Figs 1–5) with diffuse vascular malformations and diffuse ocular melanocytosis. Eight patients had ocular melanocytosis for 360° involving the episclera, iris, anterior chamber angle, and choroid in one or both eyes. Three of these cases (2, 3, 5) also had episcleral vascular malformations (EVMs) for 360° and bilateral congenital glaucoma developed. One patient with unilateral congenital glaucoma (case 4) had unilateral ocular melanocytosis, whereas another (case 6) had unilateral ocular melanocytosis but unilateral ocular vascular malformation. One patient (case 1) had asymmetric ocular melanocytosis, and congenital glaucoma developed in the more involved eye and developmental glaucoma developed in the patient at the age of 8 in the less involved eye. One patient (case 3) with bilateral congenital glaucoma had greater pigmentation in the eye in which glaucoma was present at birth. One patient (case 7) had bilateral ocular melanocytosis but only partial involvement of the left eye by a vascular malformation and elevated intraocular pressure (IOP) developed at the age of 13. One patient (case 8, Figs 6, 7) had bilateral ocular melanocytosis but no vascular...
Table 2. Oculodermal Findings

<table>
<thead>
<tr>
<th>Case No.</th>
<th>PPV</th>
<th>BCVM</th>
<th>BM</th>
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<th>FM</th>
<th>EVM</th>
<th>OM</th>
<th>Glaucoma</th>
<th>Age (yrs) at Diagnosis of Glaucoma</th>
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<td>R = L</td>
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<td>OD &gt; OS</td>
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<td>R &lt; L</td>
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<td>OU</td>
<td>OS*</td>
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PPV = phakomatosis pigmentovascularis; BCVM = body cutaneous vascular malformation; BM = body melanocytosis; FCVM = facial cutaneous vascular malformation; FM = facial melanocytosis; EVM = episcleral vascular malformation; OD = at birth; OM = ocular melanocytosis; OD = right eye; OS = left eye; OU = both eyes.

* Partial involvement.
† Ocular hypertension.

malformation in either eye and glaucoma did not develop. The last patient (case 9, Fig 8) had bilateral facial and EVMs and only partial ocular melanocytosis in one eye and had normal IOP in both eyes.

The follow-up time ranged from 1.0 to 8.5 years. Systemic findings (Table 3) included hemihypertrophy of the body or face (n = 7), palatal vascular malformations (n = 7), atopic eczema (n = 6), cortical atrophy (n = 5), seizure disorder (n = 3), palatal melanocytosis (n = 3), scoliosis (n = 1), subglottic stenosis (n = 1), polycythemia vera (n = 1), and testicular hydrocele (n = 1). Family history was noncontributory in all patients.

**Discussion**

Klippel–Trenaunay syndrome is a neurocutaneous disorder consisting of a cutaneous capillary vascular malformation (nevus flammeus) associated with venous and lymphatic malformations and soft tissue or skeletal hypertrophy or both. Parkes Weber syndrome exhibits similar findings plus arteriovenous shunting. The two have been combined as KTW syndrome, which is manifested clinically by a cutaneous nevus flammeus and hemihypertrophy of the involved organs. Numerous ocular manifestations have been reported. Glaucoma has been described in patients with KTW syndrome who also had a facial nevus flammeus.

Sturge–Weber syndrome (encephalotrigeminal angiomatosis) is characterized by a vascular malformation in the facial skin and scalp, usually in the distribution of the ophthalmic and maxillary divisions of the trigeminal nerve, and in the cerebral leptomeninges. The clinical syndrome consists of a facial port-wine stain, usually unilateral, and contralateral neurologic disorders, including seizures and hemiplegia. Facial hemihypertrophy is common. Common ocular findings include hemangiomas of the lid, conjunctiva, episclera, iris, ciliary body, and choroid. Ipsilateral glaucoma occurs in up to 50% of patients in whom the capillary vascular malformation involves both the ophthalmic and maxillary divisions of the trigeminal nerve. Episcleral vascular malformations are strongly associated with the development of glaucoma. Careful examination may be necessary to detect these, because they can be obscured by thick Tenon capsule.

Although glaucoma occurs often in infancy, it may develop in childhood or early adulthood. In infants, a developmental anomaly of the anterior chamber angle has been postulated as causal. A milder angle anomaly may be found in patients in whom glaucoma develops during childhood. Adolescents usually have a scleral angioma with increased episcleral venous pressure and a normal-looking angle on gonioscopy. In adults, elevated episcleral venous pressure or premature aging of the trabecular meshwork and/or Schlemm canal has been proposed. The KTW and SW syndromes are related closely, and the combination of the two has been reported. Because their clinical and histologic findings are similar, differing only in location of the lesion and in severity of involvement, they may represent a spectrum of disease. The nevus flammeus, often misdescribed as a hemangioma, is a congenital vascular malformation with normal endothelial turnover. Histologically, it contains dilated blood vessels without endothelial cell proliferation, a diagnostic feature of hemangioma. Oculodermal melanocytosis is characterized by hyper-
Figure 1. Case 1. Bilateral facial cutaneous vascular malformations and ocular melanocytosis. Right facial hemihypertrophy.

Figure 2. Case 1. Hyperpigmented iris, multiple dark iris processes over trabecular meshwork.

Figure 3. Case 2. Superficial cutaneous telangiectasia overlapping deeper Mongolian spots.

Figure 4. Case 4. Bilateral oculodermal melanocytosis and left facial vascular malformations and hemihypertrophy.

Figure 5. Case 6. Bilateral facial vascular malformations and ocular melanocytosis.

Figure 6. Case 8. Bilateral oculodermal melanocytosis and left facial cutaneous vascular malformations and hemihypertrophy.

Figure 7. Case 8. Diffuse ocular melanocytosis and episcleral vascular malformations.

Figure 8. Case 9. Episcleral vascular malformations and incomplete ocular melanocytosis.
pigmentation of the facial skin in the distribution of the ophthalmic, maxillary, and, occasionally, the mandibular divisions of the trigeminal nerve. Histologically, the lesion contains melanocytes deep in the dermis. Approximately 30% of patients also have ocular hyperpigmentation. Common ocular findings include hyperpigmentation of the episclera, sclera, iris, trabecular meshwork, and choroid, corneal stromal melanocytosis, pigmented deposits on the anterior lens surface, dense iris processes, and poor pupillary response to pharmacologic dilation.

Glaucoma occurs in approximately 10% of patients with ODM and may be congenital, developmental, or associated with acute anterior uveitis or hyperpigmentation of the angle. Weiss and Krohn postulated that the melanocytes increase aqueous outflow resistance, resulting in melanocytic glaucoma. Melanocytic infiltration could result in elevated IOP in eyes that already have decreased aqueous outflow. Abnormal neural crest development has been proposed to account for anomalous development of the anterior chamber angle, resulting in congenital glaucoma.

Phakomatosis pigmentovascularis is a combination of the two and has been described primarily in Asian patients with extensive cutaneous vascular malformations and pigmented nevi. Histologically, the cutaneous lesion contains telangiectatic vessels and melanocytes in the mid- and deep dermis. The vascular lesions consist of a nevus flammeus, whereas dermal melanocytes form Mongolian spots, or nevus spilus. The cutaneous vascular malformations and neurologic complications are clinically similar to both KTW and SW syndromes. Several previously reported patients with PPV, and our cases 1 through 3, 7, and 8, have had large areas of nevus flammeus associated with hemihypertrophy of the limbs or face. Patients with combined ODM and KTW and/or SW also have been reported. It appears that these conditions represent a disease spectrum.

Congenital glaucoma developed in all eyes that had 360° involvement of the episclera by both vascular malformations and melanocytosis. Elevated IOP developed in all but one of our patients who had combined facial nevus flammeus, EVM, and ocular melanocytosis. Patient 9 had extensive episcleral vascular telangiectasia, but the ocular melanocytosis was partial, with a few patches of episcleral melanocytosis and incomplete iris involvement. In addition, the extent and pigmentation of the involved anterior chamber angle were much less than in the patients in whom glaucoma developed. Although the cup-to-disc ratio in the involved eye was larger and a glaucomatous process in the past could not be ruled out, an increased cup-to-disc ratio in the hyperpigmented eye has been reported previously to occur on a congenital basis in 9.8% of a large series of patients with ODM.

Similarly, glaucoma in the left eyes of patients 1 and 3, which had less ocular melanocytosis, was less severe and occurred later in life than in the right eyes. Although there was extensive ocular melanocytosis in patient 1, the EVM was limited to the inferonasal quadrant. The IOP in this eye was mildly elevated and did not lead to glaucomatous damage during the period of follow-up.

Several patients with combined nevus flammeus and dermal melanocytosis have been reported in the nonophthalmic literature. Slit-lamp examinations were not performed, and EVMs, which are less striking than are melanocytosis, usually were not mentioned. Glaucoma developed in almost all of the patients who had combined facial nevus flammeus and ocular melanocytosis, whereas it did not develop in those without facial nevus flammeus (Table 1). Only the three patients reported by Ruiz-Maldonado et al and one eye of the patient reported by Reineke et al did not have elevated IOP. However, in the absence of a complete ocular examination and follow-up, glaucoma development could not be excluded. Glaucoma did not develop in patients with ocular melanocytosis but without a facial nevus flammeus and with a facial nevus flammeus and dermal melanocytosis elsewhere. Glaucoma developed in patients only in the eyes that had the combination with bilateral ocular melanocytosis but unilateral facial nevus flammeus or bilateral facial nevus flammeus but unilateral ocular melanocytosis. More severe glaucoma developed in the more involved eye in the patients reported by Arjona with bilateral, asymmetric facial ne-
vus flammeus, and ocular melanocytosis. The patient of Ortonne et al. had bilateral OM and vascular malformations but did not have glaucoma on the side in which only the mandibular division was involved by a vascular malformation. In another retrospective study of filtration surgery in 16 patients with SW syndrome and glaucoma, 2 also had ODM.

Several systemic findings were observed in our patients. All patients but those with atopic eczema, polycythemia vera, and testicular hydrocele have been reported previously (Table 3). Although polycythemia vera and testicular hydrocele appeared to be coincidental, there was a high incidence of atopic eczema (67%). Ataxia telangiectasia, another hereditary condition with extensive cutaneous vascular dilation, also has been reported in association with atopic eczema. The common dermal vascular anomalies may be a predisposing factor. Subglottic stenosis was described previously in a single case report. The occurrence of this potentially lethal anomaly in one of our patients (case 2) supports an association with the syndrome. Cerebral or temporal lobe cortical atrophy was the most common computed tomographic scan finding among our patients, similar to previous reports. Malignant transformation is a potential complication of ODM, and multiple benign granular cell tumors have been reported in a patient with PPV. However, tumor formation did not develop in any of our patients.

Genetic transmission in these conditions still is unclear. Trisomy 21 has been reported in single patients with KTW syndrome and SW syndrome. Although there have been reports suggesting that hereditary factors may play roles in these conditions, most reported cases, including all of ours, have been sporadic.

Neural crest is a pluripotential tissue that migrates extensively before differentiating into numerous cell types, including the peripheral nervous system, arachnoid and pia mater, melanocytes, Schwann cells, neurosecretory cells, and the skeletal and connective tissue of the face and head. Dermal melanocytes in PPV, Mongolian spots, and ODM are identical and derived from aberrant migration of neural crest cells. The SW syndrome also has been suggested to be a disorder of neural crest development. Ultrastructural examination results of a nevus flammeus showed absent paurricular nerves. An immunohistochemical study using antibodies specific to nervous system-specific S-100 protein also showed a significant reduction in perivascular nerves. This developmental anomaly of neural crest-derived vasomotor nerves was postulated to account for altered sympathetic modulation of vascular tone, leading to the progressive vascular ectasia found in this disorder.

It thus appears that ODM, PPV, KTW, and SW syndromes are all disorders of neural crest cell migration and differentiation. Because the trabecular meshwork also arises from neural crest, developmental anomalies of the anterior chamber angle in these syndromes may lead to congenital glaucoma. Extensive iris processes and hyperpigmentation of the iridocorneal angle consistently found in patients with ODM and ipsilateral glaucoma might be indicative of more extensive trabecular maldevelopment. Multiple anomalous angle structures, including an absence or poor development of the scleral spur, anteriorly inserted iris root, and thickened trabecular meshwork with or without a membrane-like tissue have been reported in patients with KTW and SW syndromes and congenital glaucoma.

As stated above, congenital glaucoma can occur in both ODM and SW–KTW syndromes. Glaucoma is more common in the latter when the ophthalmic division is involved, and glaucoma from all mechanisms occurs in approximately 10% of patients with ODM. It would appear, therefore, that the vascular malformation predisposes more strongly to the development of glaucoma than the pigmented lesion. However, when both are present at birth and extensively involve the globe, congenital glaucoma, probably due to an angle malformation, appears to be common. When involvement by one or the other of the two lesions is less, elevated IOP can appear later in life. Patients with these conditions should be examined thoroughly for ocular melanocytosis and EVMs and observed carefully for the development of glaucoma.

References

15. Sullivan TJ, Clarke MP, Morin JD. The ocular manifesta-
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