Journal of Plastic, Reconstructive & Aesthetic Surgery (2008) **61**, 889–893

JPRAS
An International Journal of Surgical Reconstruction
www.JPRASurg.com

Facial port-wine stains — clinical stratification and risks of neuro-ocular involvement☆

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Received 11 July 2006; accepted 21 May 2007

KEYWORDS
Facial;
Port-wine stain;
Sturge-Weber syndrome;
Glaucoma;
Epilepsy

Summary *Background:* Port-wine stains are capillary malformations that commonly involve the skin of the head and neck region. They may affect the underlying subcutaneous tissue and bone, and extend on to adjacent mucous membrane and conjunctiva. Ipsilateral leptomeningeal and ocular choroidal involvement occurs in a small number of cases, with variable clinical manifestations.
Aim: To analyse a series of consecutive patients with facial port-wine stains referred to our Vascular Anomalies Centre to (1) stratify their clinical manifestations, and (2) identify the risks of neurological and/or ocular involvement according to topographic pattern.
Methods: Consecutive patients with facial port-wine stains were taken from our Vascular Anomalies Database 1996–2006. Port-wine stains were topographically analysed and mapped to the sensory distribution of division(s) of the trigeminal nerve, cervical plexus, and dorsal rami of the spinal nerves.
Results: 158 patients were identified. Many of these patients had extension of their facial port-wine stains or additional separate port-wine stains on their scalp, neck, trunk or limbs. Involvement of adjacent mucosa, conjunctiva, underlying soft tissue and bone was common. Fifteen patients had associated neurological and/or ocular complications. All had port-wine stains in V1 distribution. Additional involvement of V2 and/or V3, and bilaterality were common. Seven of the nine patients (78%) with port-wine stains affecting the entire V1 had neurological and/or ocular involvement. The risk of associated neurological and/or ocular disorder in a patient with partial or full V1 involvement was 26%, glaucoma and epilepsy being the most common manifestations.
Conclusions: The clinical stratification of facial port-wine stains provides a guide to patient counselling and therapeutic interventions. Port-wine stains affecting the entire V1 distribution predict strongly for underlying neurological and/or ocular disorders that require on-going ophthalmological surveillance and/or neurological management. Although the classical Sturge-Weber syndrome

☆ Parts of this work were presented at the Royal Australasian College of Surgeons' Annual Scientific Congress, Sydney, Australia, 15–19 May 2006.

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1748-6815/$ - see front matter
doi:10.1016/j.bjps.2007.05.011

encompasses a triad of clinical manifestations, incomplete forms are not uncommon. This neuro-oculo-cutaneous syndrome is believed to be a result of vascular malformations of associated structures derived from the neuroectoderm (facial skin, eye, and parieto-occipital region of the brain and leptomeninges) during the first trimester. However, the pathogenesis of port-wine stains and Sturge-Weber syndrome remains unclear.

Port-wine stains (PWS) affect 0.3% of live births.[1] They are congenital dermal capillary malformations that have a predilection for the head and neck region.[1,2] The majority of facial PWS are macular discolourations that enlarge with the growth of the child, and darken with time. A small number of cases are associated with vascular malformations of the ipsilateral leptomeninges and/or the eye.

The triad of facial PWS, parieto-occipital leptomeningeal and ocular choroidal vascular malformations constitute the classical Sturge-Weber syndrome (SWS). However, the clinical manifestations of SWS are highly variable, with absent or varying neurological and/or ocular disorders in some cases.[3] Along with neurofibromatosis type I and tuberous sclerosis, SWS is one of the most common phakomatoses (Gk. *phakos*, meaning birthmark).[4] Nonetheless, SWS is a fairly uncommon condition. Its precise incidence is unknown and the biological basis remains poorly understood.[5]

We studied a series of consecutive patients with facial PWS referred to our Vascular Anomalies Centre 1996—2006, to (1) stratify their clinical manifestations, and (2) identify the risks of neuro-ocular involvement.

Methods

Consecutive patients with facial PWS were taken from our Vascular Anomalies Database 1996—2006. PWS were topographically analysed based on clinical records and photographs obtained prior to treatment, and mapped to the sensory distribution of division(s) of the trigeminal nerve, cervical plexus, and dorsal rami of the spinal nerves.

Diagnosis of neuro-ocular involvement in facial PWS cases was based on clinical findings. Patients diagnosed with neuro-ocular disorder were jointly managed with a paediatrician and an ophthalmologist. In patients considered to be at increased risk of extracutaneous disorder, as discussed below, the assessment of intra-ocular pressure was undertaken at 6-monthly intervals until adolescence and annually thereafter. In infants and young children, intra-ocular pressure measurement was performed at induction of general anaesthesia prior to laser treatment of PWS. Patients with glaucoma underwent closer surveillance depending on progress and control of the disease. We do not carry out routine brain imaging of asymptomatic patients although many patients had already been investigated with brain CT or MRI scanning by their physicians prior to referral to our Centre.

Results

One hundred and fifty-eight patients were identified. A large number of these patients had extensions of their facial PWS or additional separate PWS on their scalp, neck, trunk or limbs (Table 1). Extracutaneous involvement of adjacent mucosa, conjunctiva, underlying soft tissue and bone was common (Table 1).

Fifteen patients had associated neurological and/or ocular complications. All 15 patients had PWS in V1 distribution, including the entire V1 dermatome in seven. The PWS was confined solely to V1 in four patients, whilst additional involvement of V2 and/or V3 was seen in the remainder. Bilateral distribution was present in seven patients.

Seven of the nine patients (78%) with PWS occupying the entire V1 had neurological and/or ocular involvement. The risk of associated neurological and/or ocular disorder in a patient with partial or full V1 involvement was 26% (Table 2).

Of the 10 patients with glaucoma, two underwent trabeculotomy, and one had a Molteno valve implant after failed medical treatment, and the remainder were satisfactorily controlled with medical therapy. Four patients had ocular choroidal vascular malformation detected on retinoscopy. Partial visual loss and/or visual field defects were observed in half of these patients (Table 2).

Focal motor seizures were the most common type of epilepsy encountered, although grand mal and absence seizures occurred in one patient each (Table 2).

Discussion

In 1879, Sturge presented to the Clinical Society of London a 6-year-old girl with a vascular anomaly on the right side of her face, buphthalmos and left-sided focal seizures. He predicted a 'port-wine mark' on the surface of the patient's right cerebral hemisphere. More than two decades later,

Table 1 Additional distribution and extracutaneous involvement of facial PWS in 158 patients

Additional distribution	
Cervical plexus	36
Dorsal rami of cervical spinal nerves	11
Truncal	5
Limbs	6
Extracutaneous involvement	
Mucosa/conjunctiva	31
Nodule formation	24
Soft tissue hypertrophy	18
Bony hypertrophy	2