3 Vascular Tumours and Malformations

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3.1 Introduction

The localized structural abnormalities that develop during vasculogenesis and angiogenesis lead to vascular anomalies. They are divided into vascular tumours and vascular malformations. General Assembly of the International Society for the Study of Vascular Anomalies (ISSVA) divides vascular anomalies into tumours (including infantile haemangiomas and congenital haemangiomas) and malformations (Elisabeth 2016; Dompmartin et al. 2016) (Tables 3.1 and 3.2).

3.2 Vascular Tumours

3.2.1 Infantile Haemangiomas

(Terms such as strawberry naevus, capillary haemangioma and cavernous haemangioma have contributed to the diagnostic confusion in the field of vascular anomalies and are best avoided.)

Infantile haemangiomas are the most common, benign, vascular tumours encountered in infancy, occurring in up to 10% of infants, more commonly in girls than boys.

The natural history is of proliferation in the first few months of life and spontaneous involution over a matter of years. Most infantile haemangiomas reach 80% of their final size by 3 months of age. If untreated, involution is complete at a median age of 3 years (Figs. 3.1, 3.2, 3.3, 3.4, 3.5, 3.6 and 3.7).

Complications and co-morbidities: The main complications of infantile haemangiomas are ulceration, disfigurement and functional impairment.

Investigations: Investigation is rarely required as the diagnosis is usually clinical.

Occasionally ultrasound may be required:

- To distinguish infantile haemangiomas from other soft tissue masses or vascular malformations
- For plaque-type infantile haemangiomas on the face and lower trunk and before treatment with β-blockers

Prognosis: The prognosis is excellent:

- Without treatment for small infantile haemangiomas
- Larger haemangiomas if there is no functional impairment
- If it is not at an aesthetically important site
- If appropriate treatment is started in a timely fashion for infantile haemangiomas causing (or likely to cause) functional or aesthetic impairment (Higgins and Glover 2016)

The clinical photographs in this chapter are photographed by Dr. Ranthilaka R. Ranawaka, consultant dermatologist, General Hospital Kalutara, Sri Lanka.

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Table 3.1 Classification of vascular anomalies (Higgins and Glover 2016)

Simplified classification of benign vascular tumours	Classification of vascular malformations	
Infantile haemangioma (IH) Hepatic haemangioma (HH) Multifocal IH Without systemic involvement With systemic involvement **Congenital haemangiomas** Rapidly involuting (RICH) Non-involuting (NICH) Partially involuting (PICH) Tufted angioma Spindle cell haemangioma Epithelioid haemangioma Pyogenic granuloma	**Low/slow flow *capillary malformation (CM)*** Salmon patch Port-wine stain Naevus anaemicus Syndromes with CM Sturge-Weber syndrome Cutis marmorata telangiectatica congenita (CMTC) Macrocephaly-CMTC Adams-Oliver syndrome Phakomatosis pigmentovascularis Beckwith-Wiedemann Thrombocytopenia absent radii syndrome Roberts syndrome Rubinstein-Taybi syndrome Cardio-facio-cutaneous syndrome ***Venous malformation (VM)*** Syndromes with VM Maffucci's syndrome Blue rubber bleb naevus syndrome Glomuvenous malformation Cerebral cavernous malformation Verrucous haemangioma ***Lymphatic malformation** (LM – previously lymphangioma)* (Chap. 32) Microcystic and macrocystic Syndromes with LM and Gorham's disease	**High/fast flow** *Arteriovenous malformation (AVM)* Syndromes with AVM: Parkes-Weber syndrome Wyburn-Mason syndrome Cobb syndrome **Mixed/complex** *Any combination of CM/VM/AVM/LM* Klippel-Trenaunay syndrome Proteus syndrome Hereditary neurocutaneous angiomatosis CM-AVM syndrome Divry-Bogaert **Others** Hereditary haemorrhagic telangiectasia/Osler-Weber-Rendu syndrome Ataxia telangiectasia

Table 3.2 Distinction between infantile haemangiomas and vascular malformations

	Infantile haemangioma	Vascular malformation
Clinical features	Usually evident within the first week of life. Proliferate rapidly, involute over years	Usually present at birth. Proportionate growth. Do not involute
Epidemiology	More common in girls and low birth weight infants	No gender or birth weight bias
Immunohistochemistry	GLUT-1 positive	GLUT-1 negative

GLUT- 1, glucose transporter protein 1

Management: Treatment will depend on the location, morphology and stage of evolution, impact on function, risk of disfigurement and co-morbidities (Novoa et al. 2018):

1. Propranolol is now the first-line treatment. Propranolol has been shown to induce a better and faster response than systemic steroids and is associated with fewer and less concerning adverse effects (Léaute-Labrèze et al. 2016; Zhang et al. 2017).
2. Pulsed dye laser (PDL) can be helpful for the treatment of ulcerations, telangiectases and erythema post-involution.
3. Surgery may very occasionally be required in the proliferative phase if functional impairment or ulceration cannot be managed medically. Surgical reconstruction, if indicated, may be best undertaken after 3.5 years, as further aesthetically beneficial, spontaneous improvement is unlikely to occur.
4. Embolization may have a role for life-threatening haemangiomas, particularly those leading to congestive cardiac failure that has not responded to medical therapy.

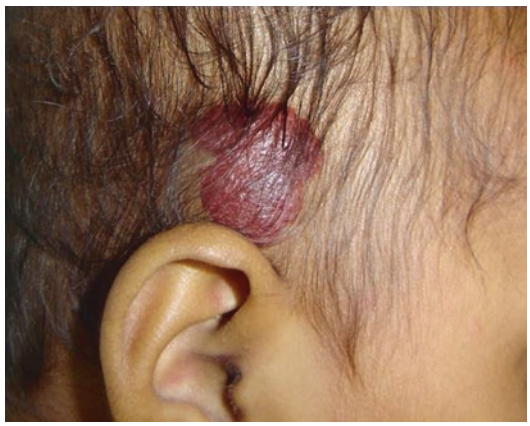

Fig. 3.1 A small capillary haemangioma in the proliferative stage on the temporal scalp of a 4-month-old infant

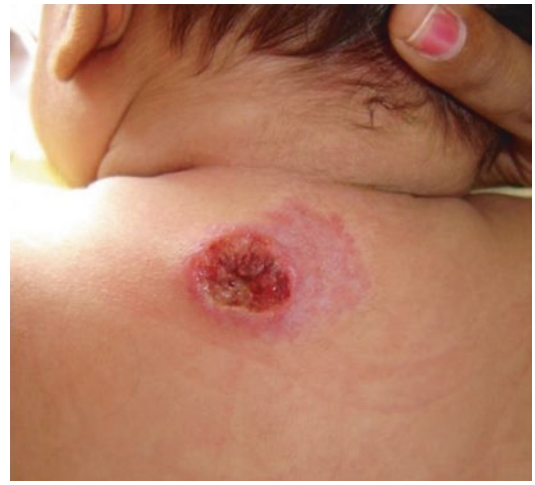

Fig. 3.2 Ulcerated superficial infantile haemangioma

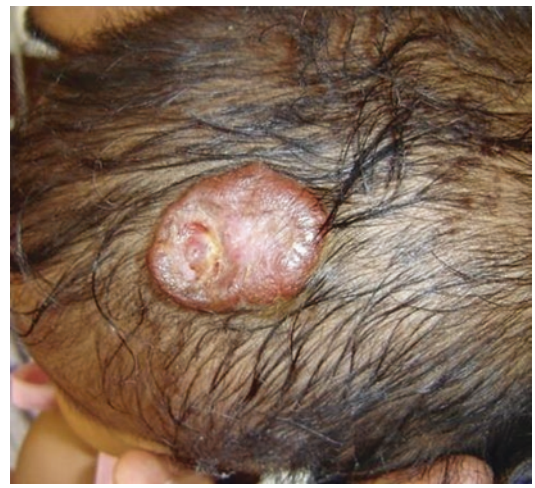

Fig. 3.3 Ulcerated plaque-like infantile haemangioma on the scalp. Ultrasound scan was done in this case to exclude intracranial extension

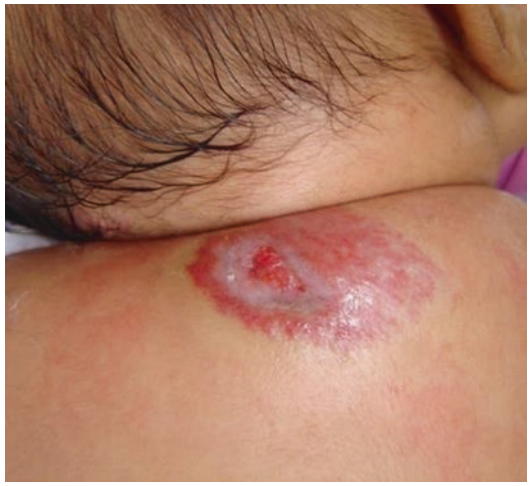

Fig. 3.4 Ulcerated infantile haemangioma in a 2-month-old baby

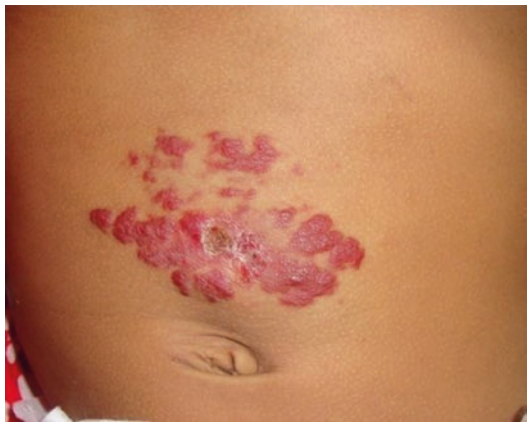

Fig. 3.5 Ulcerated infantile haemangioma on the abdomen in a 4-month-old baby

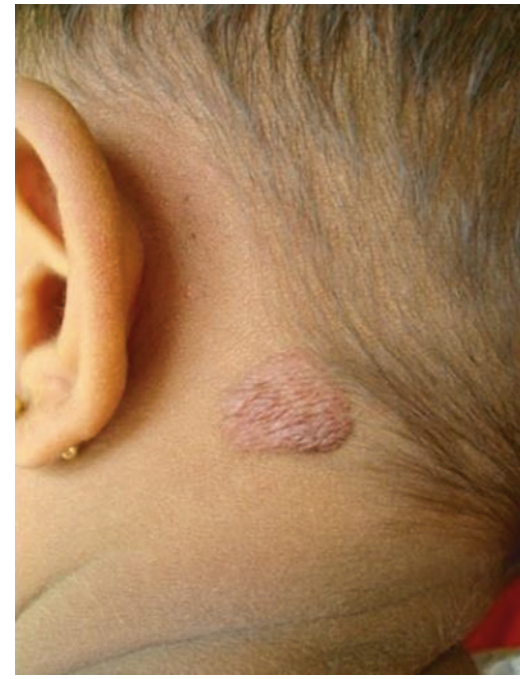

Fig. 3.6 Spontaneously resolving infantile haemangioma in a 2-year-old child

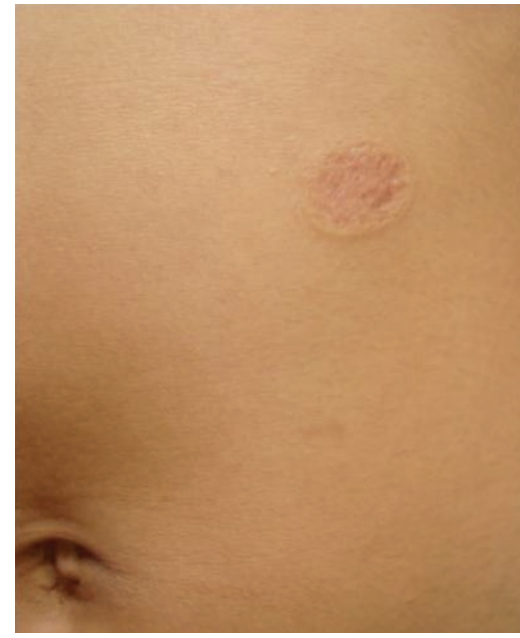

Fig. 3.7 Spontaneously resolved infantile haemangioma in a 9-year-old girl. Note residual skin changes

3.2.2 Non-involuting Congenital Haemangioma (Figs. 3.8 and 3.9)

3.2.3 Angiokeratoma of Fordyce: Penile, Scrotal and Vulval Skin

These are small benign cutaneous vascular lesions which present as red/blue or purple papules (Figs. 3.10 and 3.11).

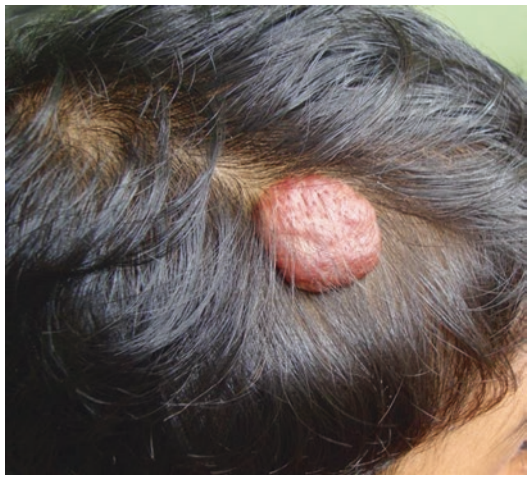

Fig. 3.8 Non-involuting congenital haemangioma in a 1.5-year-old child

3.2.3.1 Management

None is needed if the lesions are asymptomatic. If there is diagnostic difficulty, they can be surgically excised. Other treatment modalities are hyfrecation for superficial and small lesions, curettage, cautery and cryotherapy. Different lasers have been used (argon, carbon dioxide, erbium), but the KTP laser or 800-nm diode laser appears to be both effective and the least scarring, thus giving the best cosmetic result (Goldsmith 2016; Sadowsky et al. 2019).

3.2.4 Cherry Angiomas

These are very common, asymptomatic, cherry red papules seen in the skin due to abnormal vascular proliferations. They are entirely benign and increase in numbers with age.

Management: No treatment is needed in most cases. For lesions that are bleeding or cosmetically troubling, treatment options include shave excision, hyfrecation, cryotherapy and pulsed dye laser and intense pulsed light (Goldsmith 2016) (Figs. 3.12 and 3.13).

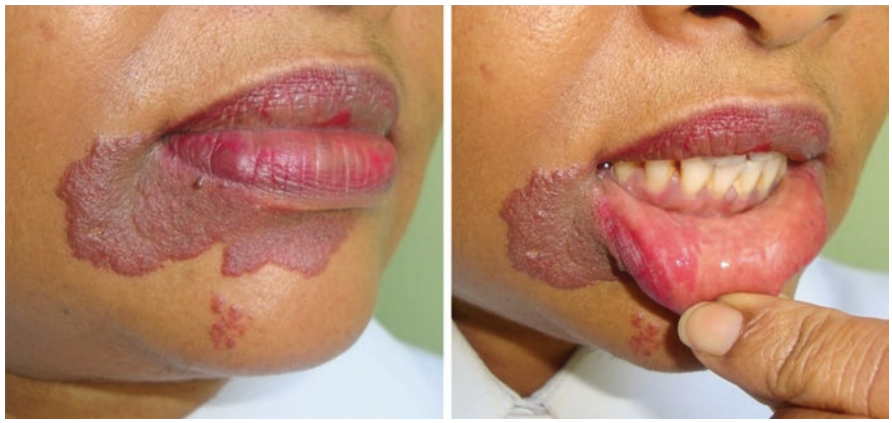

Fig. 3.9 Non-involuting congenital haemangioma in an adult woman showing cutaneous and mucosal involvement

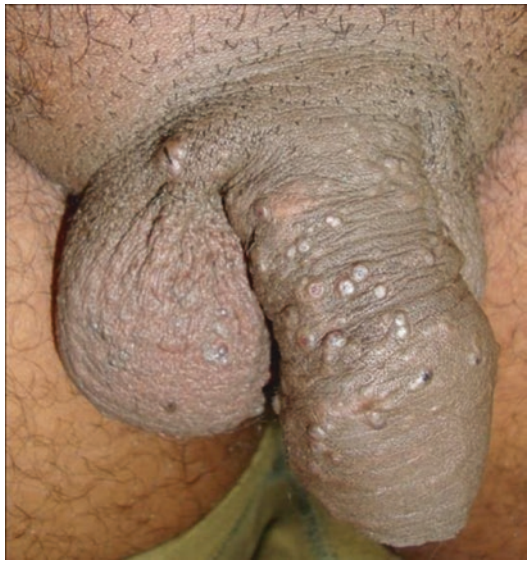

Fig. 3.10 Angiokeratoma of Fordyce in penile and scrotal skin

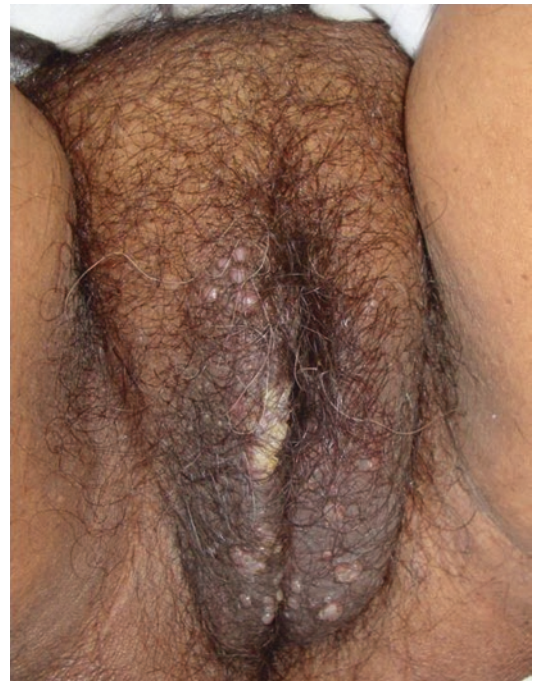

Fig. 3.11 Angiokeratoma of Fordyce in vulval skin

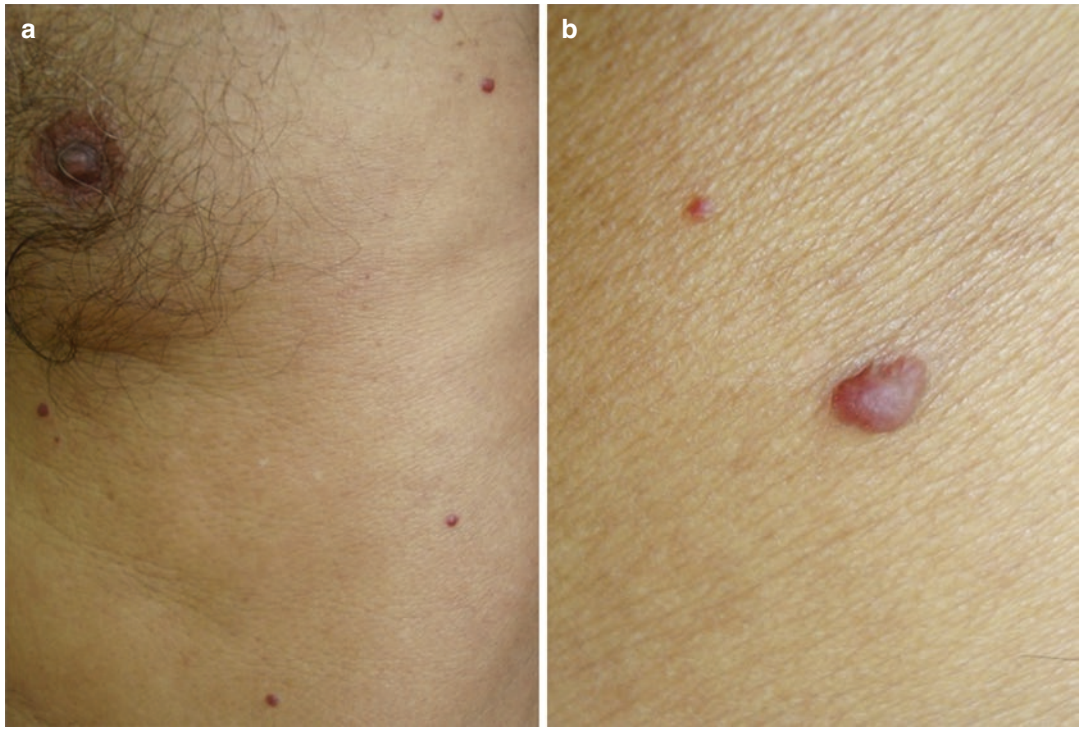

Fig. 3.12 (**a**) Multiple cherry angiomas in a 75-year-old man, (**b**) closer view

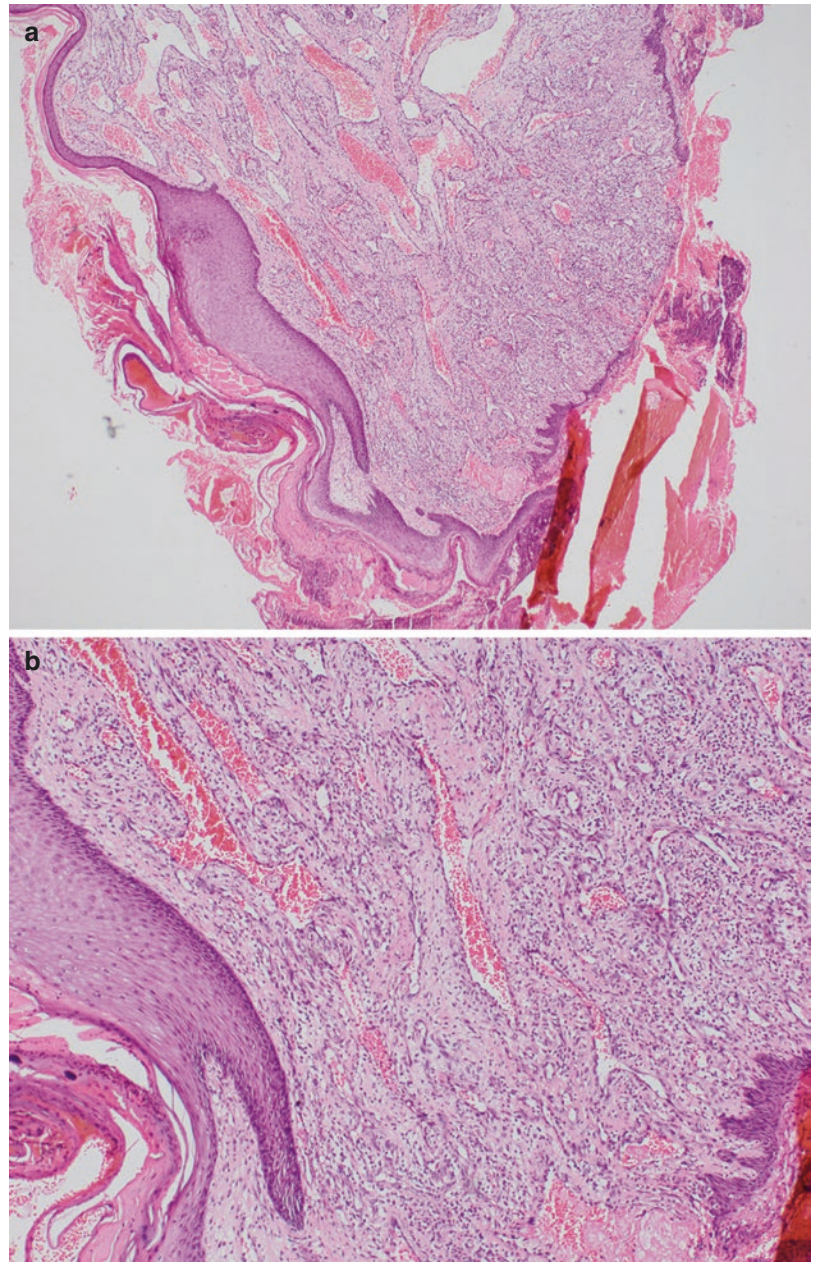

Fig. 3.13 (**a**) Haemangioma showing dilated thin-walled vascular spaces H & E stain X 40 (**b**) Haemangioma closer view H & E stain X 100 (picture courtesy Dr. Priyanka H. Abeygunasekara, Consultant Pathologist, National Cancer Institute Maharagama, Sri Lanka)

3.2.5 Pyogenic Granuloma/Lobular Capillary Haemangioma

Pyogenic granuloma is a vascular nodule that develops rapidly, often at the site of a recent injury. The lesions are very common. Spontaneous disappearance is rare. Lesions are not painful; patients mainly complain of the appearance or of recurrent bleeding (Figs. 3.14, 3.15 and 3.16).

Differential diagnosis: Keratoacanthoma and other epithelial neoplasms, inflamed seborrhoeic keratoses, melanocytic naevi, melanoma and Spitz nevi (Ranawaka et al. 2014), viral warts, molluscum contagiosum, angioma, glomus

tumour, eccrine poroma, Kaposi sarcoma and metastatic carcinoma.

Management of Pyogenic Granuloma: Simple excision is the treatment of choice, as lesions do not regress spontaneously. Local recurrence may be seen after incomplete excision (Calonje 2016). The pedunculated lesions are easy to treat by curettage with cauterization or diathermy coagulation of the base. Other treatment modalities that have been used include Nd:YAG laser, cryosurgery, intralesional steroids, flash lamp pulsed dye laser, sclerotherapy with sodium tetradecyl sulphate and even injection of absolute ethanol (Wollina et al. 2017; Plachouri and Georgiou 2019).

3.3 Vascular Malformations

Vascular malformations are localized defects of vascular development without evidence of endothelial cell proliferation, which are present at birth and never regress, grow proportionately, and have no gender or birth weight bias. They are commonly classified according to which type of anomalous vessel is involved and their flow characteristics. Vascular malformations may occur on any part of the body and may extend into deep visceral locations. They may be localized or diffuse or occur as part of a syndrome (Goldsmith 2016).

3.3.1 Capillary Malformations

There are two common types of capillary malformation with a macular erythematous patch, the salmon patch and port-wine stain.

3.3.1.1 Salmon Patch

Salmon patches are extremely common vascular anomalies and are found in all types of races. Their prevalence varies from 20 to 60%.

They present clinically with irregular pink/red macular patches with or without fine telangiectasia. The commonly affected sites are the nape of the neck extending up to the occipital scalp. The natural history of these anomalies is that the majority of the facial stains will fade within the first year of life, whereas the other sites may persist into adulthood. The sacral lesions may be associated with occult spinal dysraphism (Monteagudo et al. 2011; Leung and Telmesani 1989) (Fig. 3.17).

3.3.1.2 Port-Wine Stain

Port-wine stain is the most common vascular malformation. It presents at birth in approximately 3 of 1000 newborns and has equal sex distribution.

Clinical features: Pink, red, or purple flat lesions of variable size and with geographic borders. Usually located on the head and neck but can also be seen on the trunk or limbs. This grows proportionately with the child and persists throughout life. The lesion darkens and often thickens with age, and the underlying tissues (skin, fat, muscle and bone) can be hypertrophic (Minkis et al. 2009). This is asymptomatic but can generate important psychosocial distress (Dompmartin et al. 2016) (Figs. 3.18, 3.19, 3.20 and 3.21).

Investigations: Most do not require any investigation. Associated signs and symptoms, large size and multifocality evoke a possible 'syndromic' form.

Management of Port-Wine stain

Pulsed dye laser is the first-line treatment. It reduces coloration in 75% of patients without

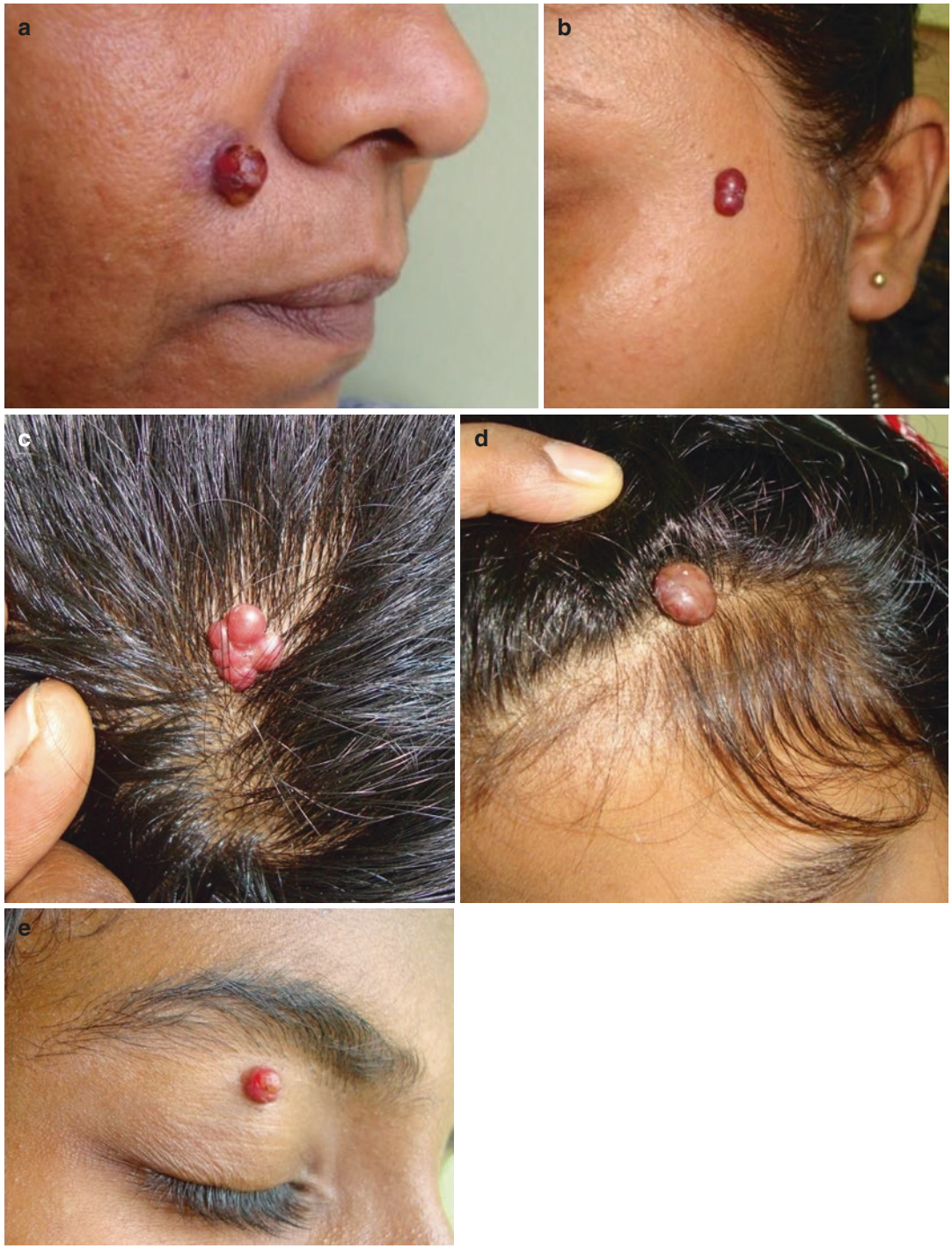

Fig. 3.14 (**a**–**e**) Different clinical presentations of pyogenic granulomas

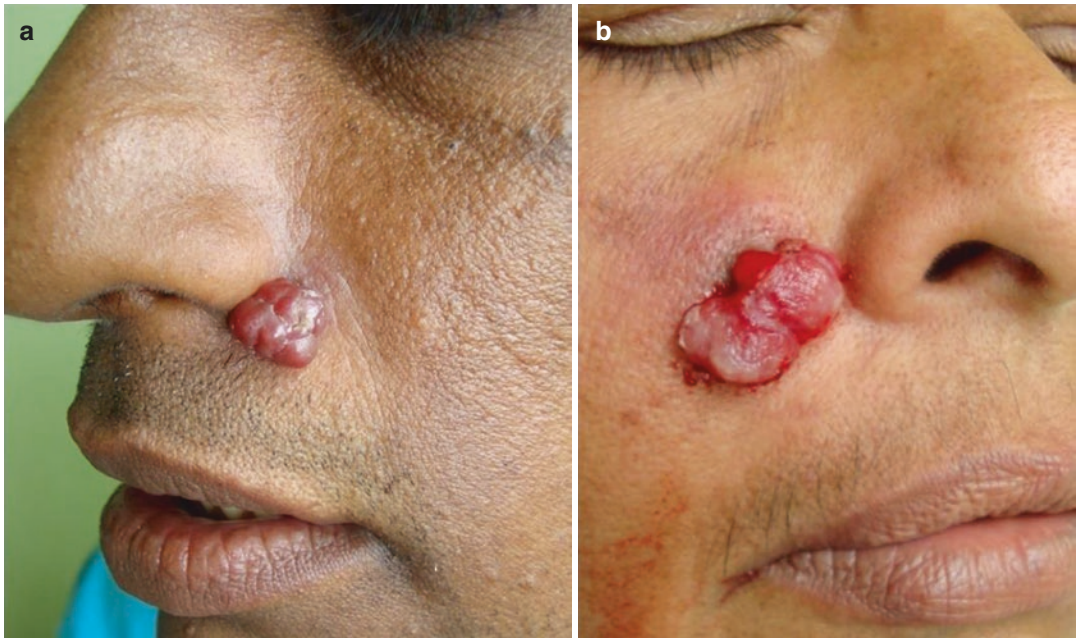

Fig. 3.15 (**a**, **b**) Pyogenic granulomas at this site mimics BCC

modifying skin texture. Laser treatment consists of several consecutive sessions. Due to pain, general anaesthesia is often used for children (Updyke and Khachemoune 2017; Savas et al. 2013).

3.3.2 Syndromes with Capillary Malformations

3.3.2.1 Sturge-Weber Syndrome

Sturge-Weber syndrome is characterized by a facial port-wine stain, leptomeningeal angiomatosis and glaucoma. The facial port-wine stain can be bilateral and/or more extensive, covering the territory of the maxillary (V2) and mandibular (V3) branches of the trigeminal nerve and sometimes the trunk and the limbs (Higueros et al. 2017; Dompmartin et al. 2016).

Clinical features: About 75% of children with intracranial vascular anomaly develop seizures, most often before the age of 2 years, with a risk of contralateral neurological deficit and learning difficulties. Gyral calcifications can be observed. The major ocular complication is glaucoma, occurring in more than 50% of patients (Higueros et al. 2017).

Investigations in Sturge-Weber Syndrome

- Brain magnetic resonance imaging (MRI)
- Ophthalmological evaluation of the fundus of the eye
- Regular assessment of intraocular pressure

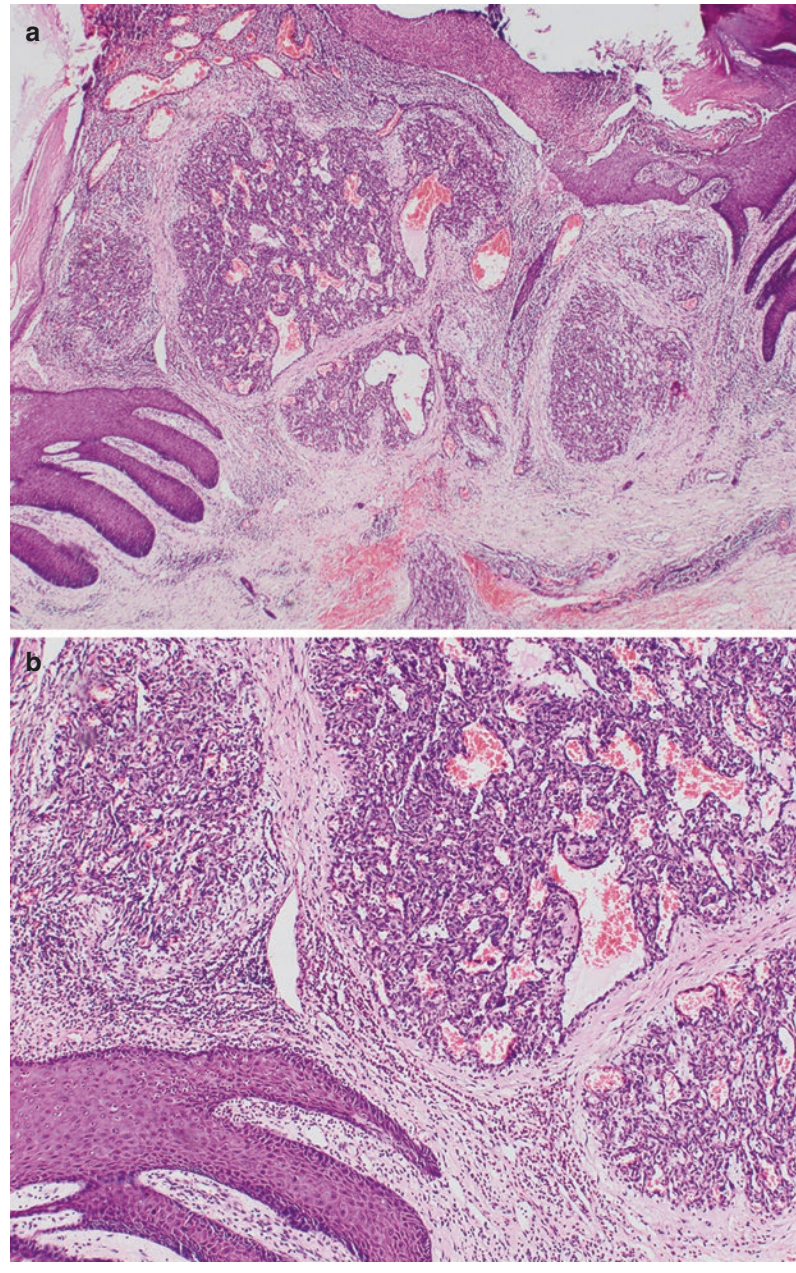

Fig. 3.16 (**a**) Pyogenic granuloma H & EX40. Lobular proliferation of small blood vessels in a myxoid stroma (**b**) Pyogenic granuloma H & E X 100 (picture courtesy Dr. Anuruddha Galapththige, Consultant Pathologist, General Hospital Kalutara, Sri Lanka)

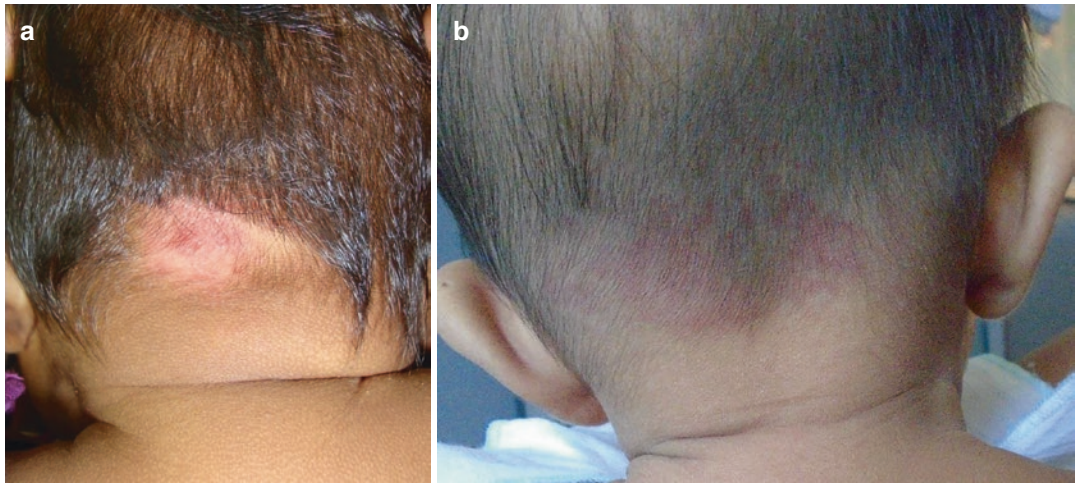

Fig. 3.17 (**a**, **b**) Salmon patch in infants at the nape of the neck extending up to the occipital scalp (photographed by Dr. Ranthilaka R Ranawaka and Dr. Maduranga Mendis, medical officer, neonatology unit, General Hospital Kalutara, Sri Lanka)

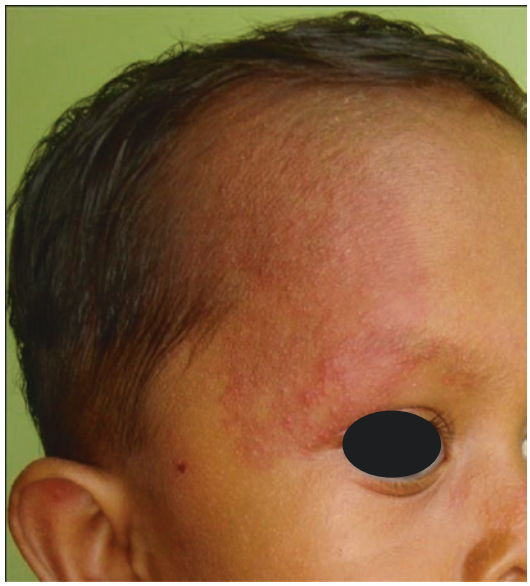

Fig. 3.18 Port-wine stain in a 5-year-old child

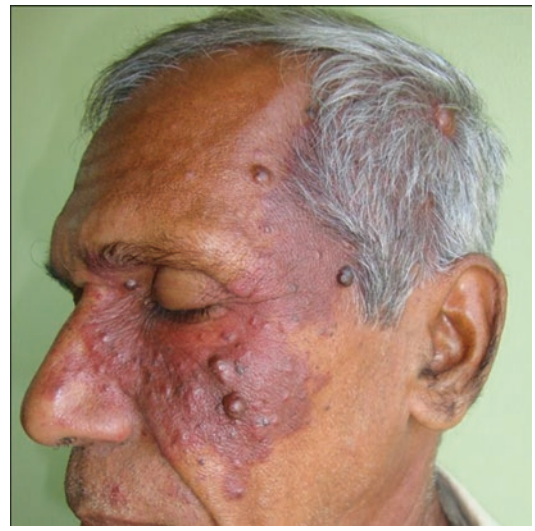

Fig. 3.19 Port-wine stain in a 74-year-old man; papular nodular eruption appeared later in life due to localized vascular proliferation. This can be prevented by early pulsed dye laser therapy

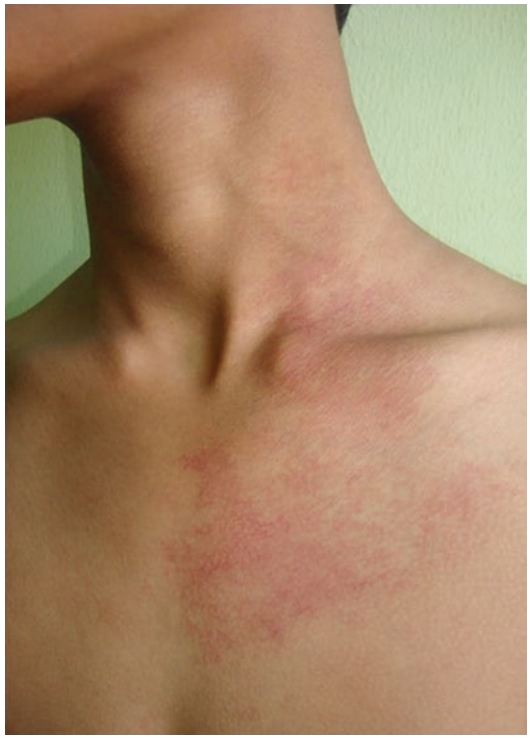

Fig. 3.20 Port-wine stain in a 17-year-old boy. This is usually located on the head and neck but can also be seen on the trunk or limbs

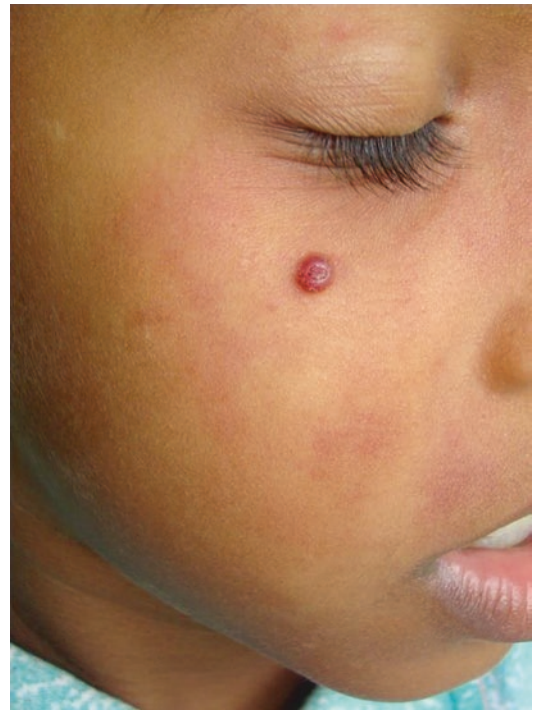

Fig. 3.21 Port-wine stain in an 11-year-old child with recently developed nodular haemangioma

Management: Pulsed dye laser

- Management of epilepsy and glaucoma is an emergency
- Intractable seizure surgery (lobectomy)
- Regular ophthalmological follow-up

3.3.2.2 Klippel-Trenaunay-Weber Syndrome

This is an uncommon syndrome characterized by overgrowth of a limb associated with a high flow capillary AVM or capillary-lymphatic AVM with multiple AV fistulae along the affected limb and increasing limb length and girth (Fig. 3.22).

3.3.3 Arteriovenous malformations (AVMs)

AVMs are the lesions with direct communications between endothelial-lined artery/arteries and vein/veins bypassing the capillary bed. AVMs are present at birth or in congenital. Acquired AVMs occur later in life due to hormonal changes or trauma. Hemorrhage and disfigurement are common reasons requiring intervention. An incomplete resection frequently leads to a recurrence of the lesion (Kolarkodi and Alnafisah 2020) (Fig. 3.23).

3.3.4 Lymphatic Malformations

3.3.4.1 Lymphangioma Circumscriptum

Lymphangioma circumscriptum is lymphatic malformations which are focal lesions composed of dilated lymphatic channels disconnected from the lymphatic system. They are mostly located on the head and neck. They are ill-defined vesicular plaques, which often invade adjacent structures. The skin can be normal in colour but becomes blue or purple when intracystic bleeding occurs (Dompmartin et al. 2016).

Recurrent erysipelas or cellulitis is major complications.

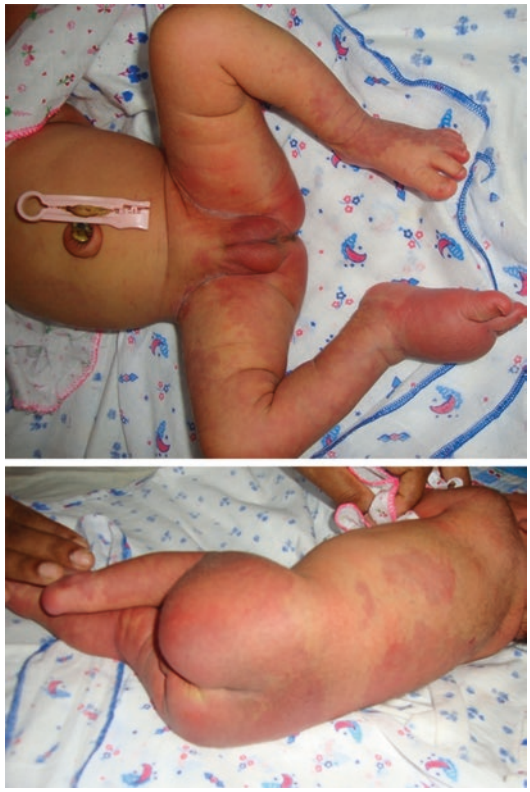

Fig. 3.22 Klippel-Trenaunay-Weber syndrome. This newborn has AV malformation, leg length discrepancy and lymphoedema (picture courtesy of Dr. Sandya Doluweera, consultant neonatologist, General Hospital Kalutara, Sri Lanka)

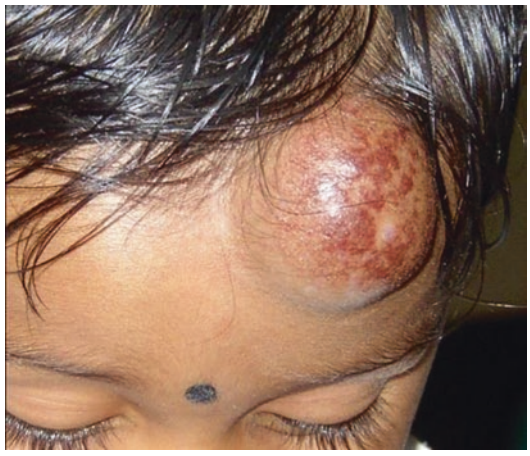

Fig. 3.23 AV malformation in a 2-year-old child (picture courtesy Dr. Kanishka de Silva MS FRCS, Consultant Oncological Surgeon, National Cancer Institute Maharagama, Sri Lanka)

Management: Multi-injection sclerotherapy, especially using bleomycin, is efficient. Laser by experienced physicians (Figs. 3.24, 3.25 and 3.26).

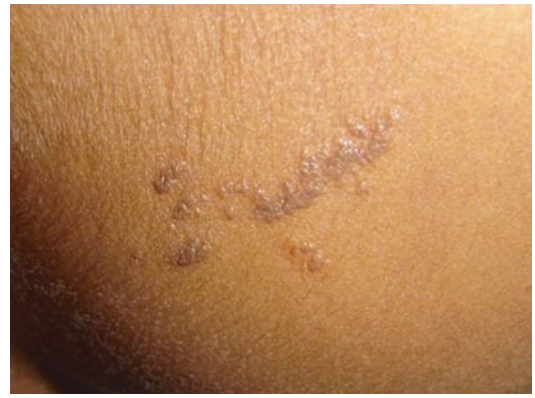

Fig. 3.24 Lymphangioma circumscriptum

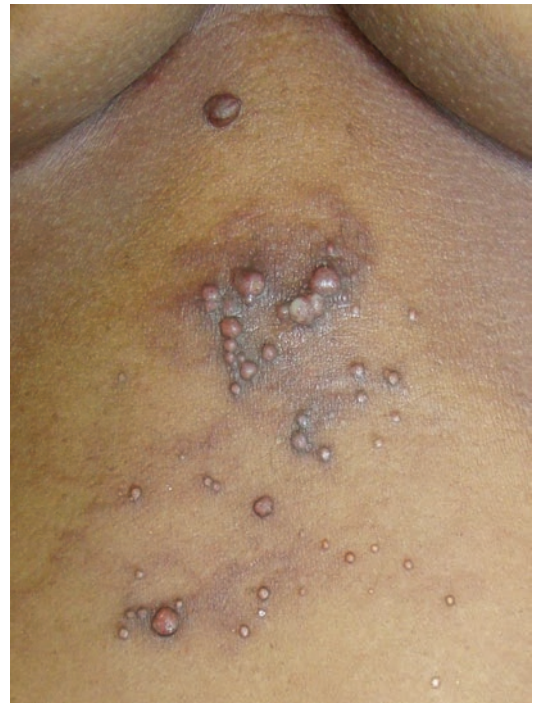

Fig. 3.25 Lymphangioma circumscriptum on the abdomen of a 40-year-old woman

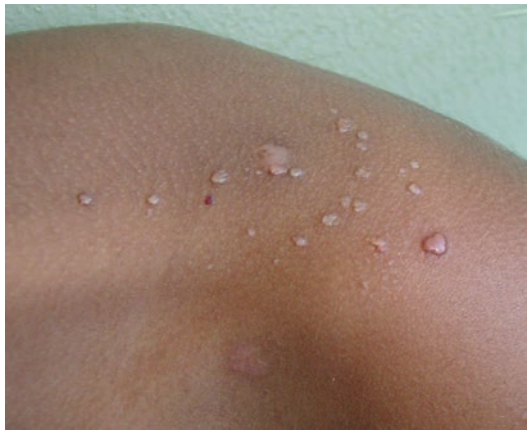

Fig. 3.26 Lymphangioma circumscriptum on the shoulder of a 7-year-old child

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