

Case 2

Current 6 year-old girl who presented first year of life with right lateral leg staining

Imaging at 2 years of age demonstrated marginal vein



What is the proper diagnosis?

Treatment strategies

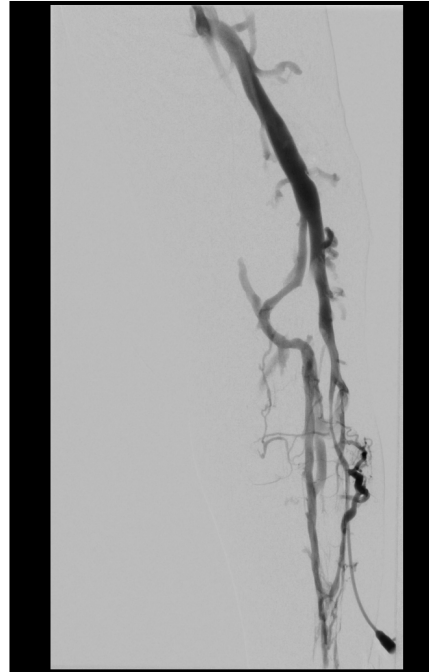
Supportive, not curative

In this patient, early identification of marginal vein made for easy interventional treatment via embolization and sclerotherapy

Management options for bothersome lymphatic component

- Sirolimus
- Laser therapy
- Surgical debulking

Remnant of Embryonic Vessels



Treatment: Medical, Interventional, Surgical

Several rounds of percutaneous sclerotherapy for painful, bleeding vesicles

Topical sirolimus trial - no relief

Oral sirolimus trial - unable due to social factors



Sirolimus (Rapamycin)

- Inhibitor of the mammalian target of rapamycin (mTOR)
- mTOR is overexpressed in KHE
- Acts in KHE to prevent downstream protein synthesis, angiogenesis and subsequent cell proliferation.

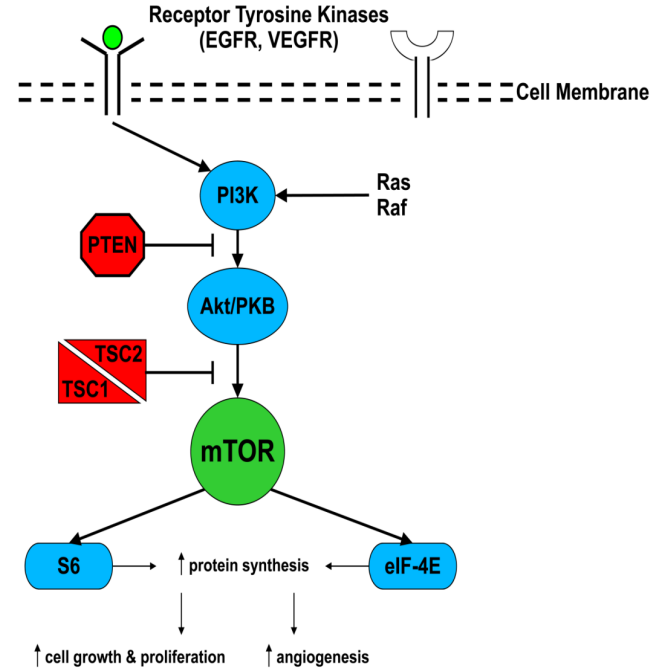
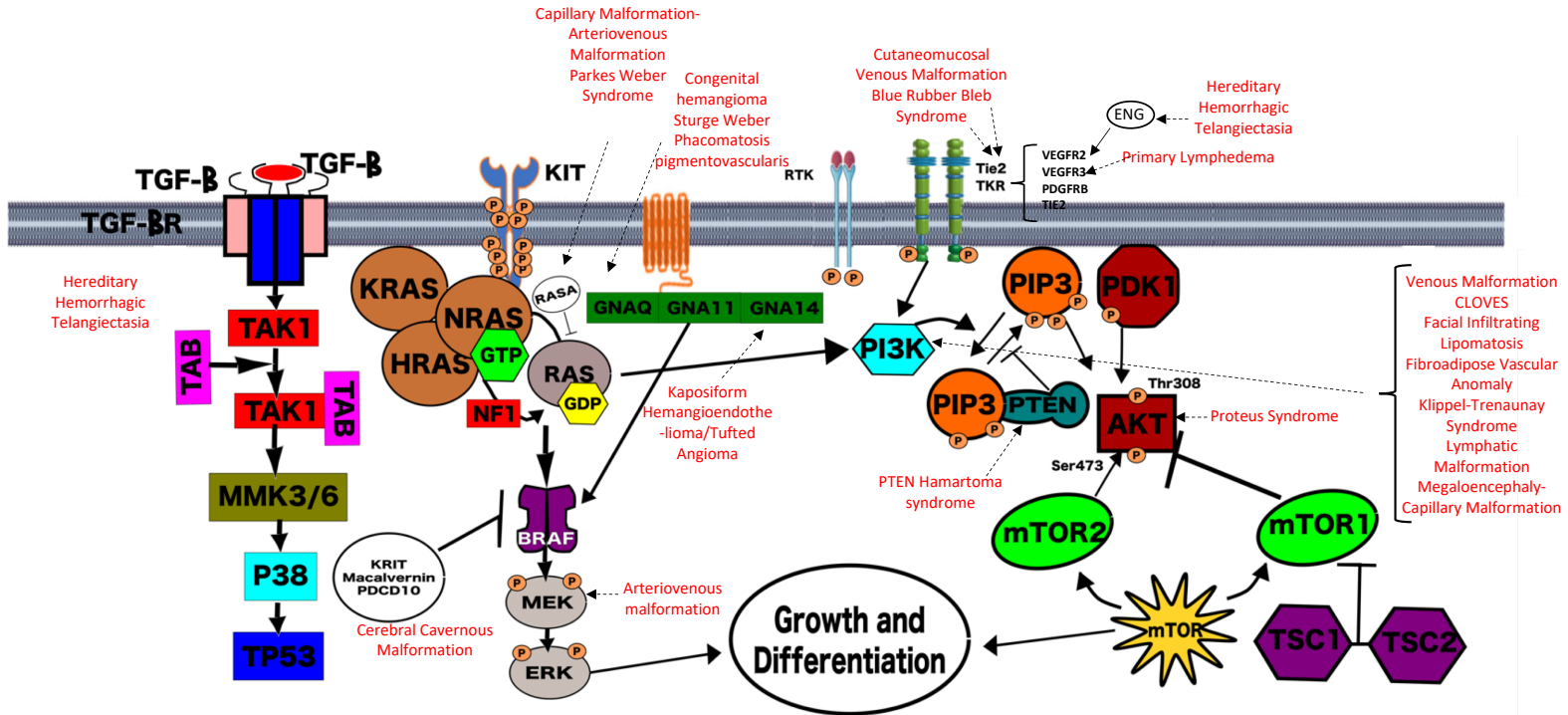


Figure 1. Genetic Pathways Implicated in Vascular Anomalies



Treatment: Medical, Interventional, Surgical

Several rounds of percutaneous sclerotherapy for painful, bleeding vesicles

Topical sirolimus trial - no relief

Oral sirolimus trial - unable due to social factors

Decision to move forward with staged surgical debulking





2 weeks post-op



3 weeks post-op

Ongoing management

Referral for wound care

- Predictable wound healing difficulties due to underlying disease

Continued discussion regarding further resection

Continue with compression

Monitor limb length discrepancy until puberty