Klippel – Trenaunay syndrome: clinical case

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Conflict of interest

I have no conflict of interest
Case presentation

- Male aged 31
- Since birth abnormal position of right foot
- Some nevi on right thigh and calf
- At 5 years, right foot fracture
- Progressive development of dilated veins on right thigh and calf with swelling
- At 21 years, operated for varicose veins on right limb
Patient complaint

- **Painful areas in different part of the limb**
- **Repeated crisis of pain in a specific area**
- **Bleeding from angiocheratomas**
- **Difficult walk by impossibility to extend knee because of foot position**
How to approach?

- Clinical evaluation?
- Angiography?
- CT?
- Doppler?
- MR?
- Angioscintigraphy total body?
Diagnostic process

1. Clinical examination
2. Duplex scan
3. MR
4. Lymphoscintigraphy
Clinical condition

- Slight right limb elongation
- Several nevi on the limb with small angiokeratomas on tight and knee
- Diffuse, dilated veins all over the limb
- Compressible areas with slow filling, some painful
- Compact, less compressible calf
- Foot blocked in extension
- No abnormal pulsation
Pain

Bleeding

D-dimer: 1288 (n.v. > 500)
Duplex scan

- Normal arteries
- Normal deep veins; no reflux
- Superficial low flow vascular masses with an extended area around the knee
- Hugh marginal vein with reflux and signs of partial thrombosis
- Combination of lymphatic and venous malformations on calf
According to Duplex Scan data, MR is performed.

Diffuse venous infiltration, intra and extramuscular

Calf
Lymphoscintigraphy

Deep

Superficial

Slower outflow right

Abnormal lateral outflow
Diagnostic conclusion

- Diffuse, infiltrating venous malformation of the whole right limb
- Some lymphatic dysplasia, truncular and extratruncular
- Large marginal vein
- Retraction of calf muscles
Treatment options

- **Conservative: elastic stockings?**
- **Sclerosis?**
- **Surgery?**
- **Laser treatment?**
- **What about blocked foot?**
Treatment decision

1. Surgical removal of marginal vein in 2 steps
2. Surgical removal of painful subcutaneous vascular areas
3. Partial skin resection including angiokeratomas
4. Elongation of achilles tendon to release foot
5. Intramuscular malformations treatable only if symptomatic by alcohol
Resection of marginal vein

Thrombous inside the vein

Resection of painful areas

Small thrombosed veins with fat hypertrophy
Result of treatment in 5 steps

- Dramatic reduction of pain
- Some slight episodes of pain on the calf
Treatment of foot block

- Surgical elongation of achilles tendon
Conclusions

- Complex, diffuse venous extratruncular infiltrating malformation of the whole limb combined with lymphatic dysplasia
- After complete diagnosis, treatment was possible
- Result was a significant improvement
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