Histology and treatment of glomuvenous malformations

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Glomuvenous malformations

Old terminology:
GLOMANGIOMA

Ectatic venous-like channels
Glomus cells

Brouillard et al, Am J Hum Genet 2002

> 70%

Brouillard et al, Am J Hum Genet 2002

Reduced penetrance and variable expressivity

Mutation in GLOMULIN

Due to somatic second-hit?

Boon et al, Hum Mol Genet 1994

Inherited
Somatic
VM versus GVM: clinical criteria

1600 patients studied

VM
- Aspect: flat, can be emptied by compression
- Color: blue
- Localization: often invades
- Pain: upon awakening, after activity, with hormonal changes
- Tx: elastic garments, sclerotherapy

GVM
- Aspect: nodular, cobblestone & plaque-like cannot be emptied
- Color: pink to purple-dark blue
- Localization: commonly only skin and subcutis
- Pain: upon awakening, after activity, with hormonal changes
- Tx: no elastic garments, surgery

Glomuvenous malformations

Nodular or cobblestone type

Plaque-like type
Glomuvenous malformations

Cutaneous and subcutaneous
Rarely superficial muscle involvement

Pleural effusion

Surgery for VM versus GVM

1 an postop

Surgery for VM

CONCLUSIONS

* GVM is a venous malformation with glomus cells
* Inherited as an autosomal dominant pattern
* Important to differentiate it from VM
* Treatment: surgery

Thank you!

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