Klippel - Trenaunay Syndrome (KTS)
When and What to do?

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Disclosure

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I have the following potential conflicts of interest to report:

☐ Consulting
☐ Employment in industry
☐ Stockholder of a healthcare company
☐ Owner of a healthcare company
☐ Other(s)

☒ I do not have any potential conflict of interest
Klippel - Trenaunay Syndrome (KTS)

In 1900, Klippel and Trenaunay first described capillary hemangiomas in one or more extremities as a congenital syndrome characterized by varicose veins, soft tissue and bone hypertrophy.

[1]
Klippel - Trenaunay Syndrome (KTS)

The incidence of this syndrome is equal in men and women.

Whether it is hereditary transition is unknown

BUT

Autosomal dominant inheritance was observed in some cases. [2]
Most, if not all, cases of KTS are caused by somatic mutations in the PIK3CA gene.

PIK3CA-related overgrowth spectrum (PROS)
KTS

Capillary malformations: 98%
Varicose veins – venous malformation: 72%
Hypertrophy: 67%

[3]
<table>
<thead>
<tr>
<th>symptoms</th>
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<tr>
<td>Capillary malformation</td>
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<td>Varicose vein</td>
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<td>Hypertrophy</td>
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<td>Popliteal vein Agenesis-Anomaly</td>
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<td>Bone malformation</td>
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<td>Port wine stain</td>
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Capillary malformations

This is the most common cutaneous manifestation of KTS. Typically, capillary malformations involve the enlarged limb.

It is seen 98% of patients.

Hypertrophy, hemorrhage, and color changes in the extremities are often the first conspicuous in patients due to venous malformation and varicosities at the skin.
Varicose veins - Venous malformations

Can occur in both the superficial and deep venous systems.

Superficial venous abnormalities:
ectasia of small veins or large venous malformations.

Deep venous abnormalities
aneurysmal dilatation, aplasia, hypoplasia, duplications, and venous incompetence.
Hypertrophy

Enlargement of the extremity:

may be caused by bone abnormality

circumferential soft-tissue

Hypertrophy or

lymphatic malformation.

Extremity growth usually stops at the end of the Adeloson period.
Port - wine stain

port-wine stain
KTS
22 y.o female
Popliteal vein agenesis
Minimal hypertrophy
Port-wine stain
55 y.o male

Port wine stain and multiple varicose veins on right leg.

Aneurysms were detected in the popliteal and femoral vein.
KTS Treatment

How can we do for this patients
And
What is the purpose of treatment in KTS
KTS

Treatment

There is no cure for KTS

Treatment is symptomatic and supportive.

Reduce symptoms and risk of complications.

Conservative treatments most effective for undesired side effects.
What can we do
What to do?

- Elastic Socks

-Conservatively

• Protects the extremity from trauma.

• Used to reduce the effects of venous insufficiency.
What to do?

-Surgery
-Ligation and Stripping

- a small incision to prevent pooling of blood.
- Ligation and striping can be used for removal of the vein.

- should be known that deep veins are normal
What to do?

Ambulatory phlebectomy

should be known that deep veins are normal
What to do?

**Sclerotherapy**

Sclerotherapy can eliminate the pain and discomfort of varicose veins. It can prevent complications such as venous hemorrhage and ulceration.
What to do?

**Sclerotherapy**

Foaming agent pushes blood in the vein so the sclerosing agent will have better contact with the vein wall.
What to do?
-Sclerotherapy

Several sclerotherapy treatments usually are required to achieve the desired results.

under ultrasound guidance
What can we do

And sometimes we can't do anything
What to do?

Sometime

Nothing

no right leg complaints

and happy

Loves his right leg more than his left.
KTS

References

Thanks...
Can it be seen in the upper extremity

Limited number of cases have been reported in the literature
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