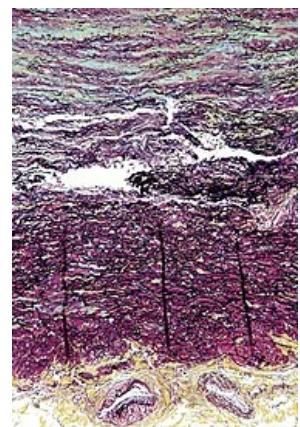


Cystic medionecrosis

Cystic medionecrosis (also called Erdheim's disease, cystic medial necrosis or cystic medial degeneration) is a disease affecting the large arteries, especially the aorta, which is characterized by **accumulation of mucopolysaccharides in the media of these vessels and** 'loss of muscle and elastic fibers. These lesions resemble cysts in appearance.^[1]

In most cases, cystic medionecrosis occurs independently, but not infrequently also within congenital diseases of the connective tissue, e.g. Marfan's syndrome, Ehlers-Danlos syndrome or annuloaortic ectasia. The cause of this disease is unknown. An **autosomal dominant** inheritance is assumed.^[2]



Despite its name, the lesions described in cystic medionecrosis are not always necrotic^[3] and do not always form cysts^[4].

Cystic medionecrosis increases the risk of developing **aneurysm** or **aortic dissection**.

Links

External links

- Cystic medionecrosis (English Wikipedia) (https://en.wikipedia.org/wiki/Familial_thoracic_aortic_aneurysm)

Related Articles

- Mucous dystrophy
- Intercellular mass disorders
- Mucopolysaccharidoses
- Inherited disorders of sugar metabolism

References

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