

A giant pulmonary artery aneurysm associated with congenital keratoglobus

Doğumsal keratoglobus ile ilişkili dev pulmoner arter anevrizması

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Dear Editor,

We read the article by Alur et al.^[1] with a great interest. We congratulate the authors and thank them for sharing their experiences with us. Congenital keratoglobus refer to as an association with connective tissue disorders such as Ehlers Danlos syndrome type VI, Marfan syndrome, and Rubinstein-Taybi syndrome.^[2] Ehler Danlos syndrome is an autosomal dominant connective tissue disorder caused by defective collagen synthesis. It is categorized by genetic mutations and clinical features. Hyperelasticity and vulnerability of the skin and joints, and friability of the eyes and vessels are the characteristic features.^[3] Keratokonus and keratoglobus, which are postulated as ‘Oculus Fragilis’, are the main features of Ehler Danlos type VI. The diagnosis is confirmed based on the abnormal synthesis of the fibroblasts type III procollagen molecules or based on the identification of genetic mutation for type III procollagen (COL3A1). Ehlers-Danlos type VI has a close relationship with a deficiency of lysyl hydroxylase in cultured fibroblasts.^[4]

However, we are unable to reach limited data on the aforementioned disorders in the pathological report of the resected tissues of this patient, except the ‘detached media layer with fibrin accumulation in the detached areas’. Did the patient have a family history and phenotype such as hypermobility or easy bruising? Did you confirm the type of collagen tissue disorder with any method such as biochemically or histopathologically?

We suggest that more detailed information about the aortic and pulmonary annular size and the patient’s body mass index would contribute to gain a better understanding the replacement reason and graft choice decisions. We believe the aforementioned details would enrich the manuscript.

Declaration of conflicting interests

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Author Reply

Dear Editor,

We read with great interest the additional commentary of the author(s) on our recent case report titled “A giant pulmonary artery aneurysm associated with congenital keratoglobus”.^[1] Our answer to the letter is given below:

The pathological examination of the resected aorta and pulmonary artery revealed that the media layer was detached with fibrin accumulation in the detached areas. However, the type of collagen tissue disease was not specified in the report.

The family history of the patient did not include any collagen tissue disease. Unfortunately, biochemical or histopathological investigation for collagen tissue disease was not available at that date in our hospital.



In addition, a contrast-enhanced thoracic computed tomography scan of the aortic root revealed a size of 55 mm; the supra-avalvular aortic diameter was 28 mm, and the pulmonary conus was dilated and 61 mm at its widest point.

The body mass index of the patient was 22.9 kg/m².

We would like to thank the authors for their contribution.

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