Klippel-Trenaunay syndrome as a rare cause of chronic thromboemboembolicpulmonary hypertension

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ABSTRACT

Klippel-Trenaunay syndrome (KTS) is a congenital disorder characterized by cutaneous capillary malformations, soft-tissue and bone hypertrophy, multiple capillary, venous or lymphatic malformations. KTS is associated with recurrent thromboembolic events that may lead to chronic thromboembolic pulmonary hypertension (CTEPH). We report five cases of KTS complicated by CTEPH. KTS was diagnosed in childhood; fourpatients had previous history of acute pulmonary embolism. Hemodynamics showed a severe disease, in accordance with the symptoms (dyspnea class III NYHA in 4 patients, class II in 1 patient). HRCT and pulmonary angiography confirmed proximal CTEPH in one patient and distal CTEPH in 4 patients. Evolution after thromboendarterectomy was good with pressure normalization, while for the rest of the patients response to specific therapy was poor (2 deaths, 2 survivors). In conclusion, CTEPH/KTS is a severe condition with a poor prognosis that requires careful monitoring of the anticoagulation therapy and specific multidisciplinary follow-up in expert centres.