

Research Journal of Pharmaceutical, Biological and Chemical Sciences, 2016, vol.7, N6, pages 2958-2970

Rendu-Osler-weber disease. Case report

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Abstract

Rendu-Osler-Weber disease is the most frequently observed genetic hemorrhagic angiopathy with local wall thinning, distention of microcirculation vessels' lumen, and incomplete local hemostasis. This pathology is inherited per autosomal-dominant type with a various pathologic gene's frequency of occurrence. As per the literature, the frequency of occurrence of this anomaly equals 1:100 000, although as of today, according to the newest studies, it is possible that the number of this nosology's events would increase up to 1:5000-8000. According to the modern ideas, Rendu-Osler-Weber disease is an example of vascular pathology (with the distress of anatomical continuity of vessels) with a congenital tendency to malformation of blood vessels (vascular dysplasia). This anomaly is characterized by the inadequacy of mesenchyme. The anatomical substrate of the disease of represented by thinning of the vessel wall with the lack of elastic and muscular layers. That's why the wall is represented by the endothelium solely, surrounded by the loose connective tissue. Because of this, arteriovenous aneurysms appear, which cause hemorrhages due to the vulnerability of vessel walls.

Keywords

Rendu-Osler-Weber disease, Vascular pathology, Vessel wall