Original Article

Clinical Manifestations Of Connective Tissue Dysplasia In Children With Glomerulonephritis

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Abstract

Glomerulonephritis is a multifactorial disease and is characterized by high rates of morbidity and disability of the child population in modern populations. Despite extensive study, the development and progression of glomerulonephritis (GL) remains one of the leading problems of nephrology. 54 children with GN, aged from 1 to 6 years, who were treated at the regional children's clinical association, were examined. Thus, the presence of multiple stigmas of DST and dysembriogenesis in a patient with GN may be an indirect criterion for predicting a severe course of the process with damage not only to the glomerular apparatus, but also to the basement membrane. In children with glomerulonephritis, connective tissue dysplasia occurs with a higher frequency than in the population of practically healthy children. The definition of stigmas of DST and dysembriogenesis is a simple, fairly informative method, publicly available and not difficult for a pediatrician.

Keywords: dysplasia, connective tissue, children, glomerulonephritis

Introduction. Glomerulonephritis is a multifactorial disease and is characterized by high rates of morbidity and disability of the child population in modern populations [1,3,8]. Despite extensive study, the development and progression of glomerulonephritis (GL) remains one of the leading problems of nephrology. In recent years, special attention has been paid to children with signs of systemic changes on the part of various organs associated with the peculiarities of metabolism and structure of connective tissue, which clinically manifest themselves as a complex of signs and are designated in the literature as connective tissue dysplasia (DST) [2,7]. The question of the role of connective tissue dysplasia in the formation of forms of glomerulonephritis resistant to therapy remains unresolved. Connective tissue dysplasia syndromes are genetically heterogeneous, and according to a number of authors [1, 2, 5,9], they are detected with high frequency in children with renal pathology, including in patients with pyelonephritis, interstitial nephritis, cystitis, nephroptosis, glomerulonephritis and other diseases [3, 4, 6].

The aim of the work was to study the clinical and laboratory features of glomerulonephritis (GL) in children with connective tissue dysplasia (DST).

Materials and methods of research. 54 children with GN, aged from 1 to 6 years, who were treated at the regional children's clinical association, were examined. In 24 patients, glomerulonephritis occurred with nephritic syndrome, in 18 — with nephrotic syndrome (NS), in 12 — with NS and hematuria. A generally accepted clinical and laboratory examination was performed in all patients, which included the identification of stigmas of DST and dysembriogenesis. Thus, the presence of multiple stigmas of CTD and disembriogenesis in a patient with AG may be an indirect criterion for predicting the severity of the process, damaging not only the glomerular apparatus but also the basement membrane. Because AG with nephrotic syndrome and AG with nephrotic syndrome were more common in

patients with multiple stigmas of CTD and dysembryogenesis, the clinical manifestations in these patients had certain characteristics.

In patients with AG with CTD stigma, edema syndrome was more common in anasarca (39.20 ± 5.49%) and in patients with moderate edema (16.46 ± 4.17%, r <0.05) and in children with AG without CTD stigma, dominated the eyelids and legs (80.00 ± 10.69%, r <0.01). The tumor persisted longer (12.32 ± 1.05 days) in patients with AG with CTD stigma, and the duration of tumor in patients without CTD stigma was 8.07 ± 1.31 days (r> 0.05). Macrohematuria is less common in patients with AG without CTD stigma (86.70 ± 9.08%, r> 0.05). Its duration does not depend on the presence or absence of CTD stigmas. Proteinuria up to 1 g / l In patients with AG without CTD stigma (73.33 ± 11.82%), proteinuria up to 2 g / l was observed in patients with AG with CTD stigma (r <0.05). The daily average rate of proteinuria was 2.6 times higher in patients with CTD stigma than in patients without CTD stigma. The degree of leukocyturia does not depend on the number of CTDs (r> 0.05).

Acute phase parameters (cialic acid, diphenylamine test (DFA), seromucoid, C-reactive protein), CIC and cryoglobulins, fibrinogen levels were significantly increased in patients with AG with CTD stigma. This is because the presence of CTD stigmas reflects some features of the metabolism of connective tissue structures, acute phase parameters, levels of CIC and cryoglobulins were analyzed depending on the presence or absence of fibrinogen CTD stigmas.

Thus, in patients with AG with CTD stigma, tsial acids were elevated in 62.75 ± 6.77% of cases, with an average rate of 261.0 ± 11.0 units; DFA increased by 70.59 ± 6.38%, the average level of DFA was 0.290 ± 0.011 units; seromucoid increased in 49.02 ± 7.00% of patients, its mean level was 0.32 ± 0.01 op.pl. n patients with AG without CTD stigma, sialic acid, DFA, and seromucoids were elevated in only 1/6 of patients, and the mean level of sialic acid was 182.5 ± 17.0 arb units, DFA - 0.210 ± 0.017 u.p. (p <0.01), seromucoid - 0.22 ± 0.02 op.pl. (r <0.001). The mean level of C-reactive protein in the presence of CTD stigmas was 4.6 times higher than in patients without CTD stigma (r <0.01). The mean fibrinogen level was slightly higher in the presence of CTD stigmas (4.40 ± 0.36 g / l, r> 0.05). The number of CTD stigmas affected the growth phase of acute phase parameters, CIC, cryoglobulins, fibrinogen. In patients with multiple CTD stigmas, all of these rates were found to be high.

Results and their discussion. In the examination of 54 children with GN, DST stigmas were detected in 81.2 ± 3.9% of patients, multiple DST stigmas (3 or more) were detected in 41.5 ± 5.0%. Of the stigmas of DST, anomalies of the hands and feet (39.4 ± 4.0%) and flat feet (35.4 ± 5.0%) were most common. Pigmented spots and hypermobility of joints were found with the same frequency (29.6 ± 4.89%), and posture disorders, scoliosis were detected in 18.2 ± 4.1% of patients. Other stigmas of DST (chest deformity, hernias, myopia, tall stature and long fingers, sandal-shaped cleft, gallbladder deformity, mitral valve prolapse) were found in less than 9% of the examined. More than 5 stigmas of DST had 4 out of 54 children. The absence of stigmas of DST was detected in 12.9 ± 3.78% of children with GN. In various forms of GN, stigmas of DST occurred with almost the same frequency (p>0.05). Three or more stigmas of DST occurred with equal frequency in patients with nephrotic syndrome (39.66 = 6.42%) and nephrotic syndrome (40.91 = 10.73%). This may indicate that the peculiarities of metabolism, immunity in children with DST predispose to the development of OGN, but do not determine its form. Since DST is genetically determined, the peculiarities of connective tissue metabolism can occur in utero, which affects the formation of certain stigmas of dysembriogenesis. Dysembriogenesis stigmas were detected in 91.5 ± 2.9% of patients with GN, multiple dysembriogenesis stigmas (3 or more) They were detected in 51.0 ± 5.2% of children, that is, in every second patient with GN. Of the stigmas of dysembriogenesis, the tendency to syndactyly of the II, III toes was most common (69.5 ± 4.2%), with the same frequency — Gothic palate and hypertelorism (56.3 ± 5.1 and 52.1 ± 5.2%, respectively), somewhat less often — deformation of the earlobes (19.8 ± 4.1%), low hair growth on the forehead (18.0 ± 4.0 %). Other stigmas of dysembriogenesis, such as skull shape anomalies, epicanthus, kidney anomalies, eye anomalies, cryptorchidism, extra nipple on the breast, hypertrichosis, were found in less than 5% of the examined children. 8 out of 54 children (14.8 ± 2.8%) had more than 5 stigmas of dysembriogenesis. The occurrence of dysembriogenesis stigmas in various forms of GN had a number of differences. In the nephrotic form of GN, 3 or more dysembriogenesis stigmas were more common than in the nephritic form, which were detected in 58.2 ± 10.2% of patients with nephrotic form and in 41.3 ± 6.5% with nephritic (p <0.05). At the same time, 1-2 dysembriogenesis stigmas were more often detected in nephritic syndrome (10 patients, 41.6 ± 4.2%) and only in 4 out of 18 patients with nephrotic syndrome (p<0.05). The presence of multiple stigmas of dysembriogenesis in patients with nephrotic syndrome indirectly indicates the negative influence of the peculiarities of connective tissue metabolism, membrane permeability, immunity, its regulatory systems (cytokines) on the occurrence of nephrotic syndrome in PURULENT. In isolated urinary syndrome, the frequency of dysembriogenesis stigmas did not differ from the frequency in OGN with nephritic syndrome, so these two groups of patients were not separated. The nature of dysembriogenesis stigmas in the groups of patients with various forms of GN did not significantly differ (p>0.05). Thus, the presence of

multiple stigmas of DST and dysembriogenesis in a patient with GN may be an indirect criterion for predicting a severe course of the process with damage not only to the glomerular apparatus, but also to the basement membrane. Since patients with multiple stigmas of DST and dysembriogenesis were more likely to have GN with nephrotic syndrome and GN with nephrotic syndrome with hematuria, clinical manifestations in these patients had certain features.

Conclusions.

1. In children with glomerulonephritis, connective tissue dysplasia occurs with a higher frequency than in the population of practically healthy children.

2. The definition of stigmas of DST and dysembriogenesis is a simple, fairly informative method, publicly available and not difficult for a pediatrician.

3. The study of the identification of stigmas of DST and dysembriogenesis is particularly relevant at the present stage due to the deterioration of the environmental situation and the change in the classical clinic of the disease.

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