Case Report

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Relapsing Polychondritis and Hematuria: A Case Report

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ABSTRACT

Relapsing polychondritis (RP) is very rare inflammatory disorder characterized by episodic, progressive, and destructive courses affecting cartilages. Renal involvement also is very rare presenting sign in these patients. It has seldom been described in children. We describe an 11-year-old girl with RP and complaint of frequent episodes of hematuria, and dysuria with fever and periorbital edema that auricular cartilage involvement appeared after renal involvement. Renal involvement in the setting of RP is mainly important, and need close observation.

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Introduction

Relapsing polychondritis (RP) is one of the systemic inflammatory condition that involve tissues containing cartilaginous structures such as nose, ear, joint, airway tract, and kidney (1). It occurs between the age of 40 to 60 years, but may occur in childhood (2). Most common clinical picture of RP is auricular involvement, and less common feature is glomerulonephritis (3, 4).

We describe an 11-year-old girl with RP that her first presentation was hematuria.

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Patient was an 11-year-old girl with the clinical setting of frequent episodes of dysuria and hematuria, intermittent fever, periorbital edema, and swelling of right ankle initiated 4-months prior to admission (Figure 1). In addition, the patient suffered from arthralgia in metacarpophalangeal (MCP) joints, and myalgia in calf muscles which lead to gait

problems. Totally, other significant problems included weight loss over last month, anorexia, and conjunctivitis accompanied with periorbital edema and erythema.

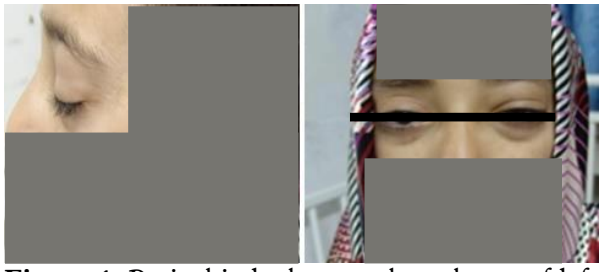

Figure 1. Periorbital edema and erythema of left eye and ear

In her physical exam, she was a conscious teen girl with visible pallor in general appearance, and left palpebral edema. In extremities, erythema, edema, and limited range of motion were detected in right ankle. Metatarsophalangeal (MTP) joints in right foot were swollen and tender.

During first days of admission, swelling, erythema, and tenderness in the superior segment of right auricle appeared (auricular chondrites). Left auricular chondrites occurred after 3-4 days.

Primary laboratory tests revealed an elevated erythrocyte sedimentation rate (ESR) of 60 mm/hour, C-reactive protein (CRP) of 57 mg/l, anemia (hemoglobin: 8.8 g/dl), and abnormal urine analysis (U/A) with persistent hematuria (3+) and proteinuria (2+). Laboratory findings of 24-hour urine were as creatinine of 500 mg, and protein of 1600 mg. Urine calcium level, and also calcium/creatinine ratio were normal.

Lupus anti-coagulant, anti-cardiolipin, anti-phospholipid A (IgG), and anti-nuclear antibody (ANA) were all reported as negative.

Thyroid function tests were normal. Purified protein derivative (PPD) reaction was 20 mm; meanwhile polymerase chain reaction (PCR) of urine and gastric aspiration specimen did not detected mycobacterium tuberculosis DNA.

Computed tomography (CT) scan revealed orbital pre-septal edema with invasion to post-septal region. Maxillary and ethmoid sinuses were normal.

Biopsy of involved cartilage revealed levels of chondrocytes degeneration, and eosinophilic discoloration and degenerative changes of background material which were suggestive of RP. Kidney needle biopsy demonstrated findings compatible with focal and segmental glomerulosclerosis, as well as mild chronic interstitial inflammatory cells infiltration, and foci of tubular atrophy (Figure 2).

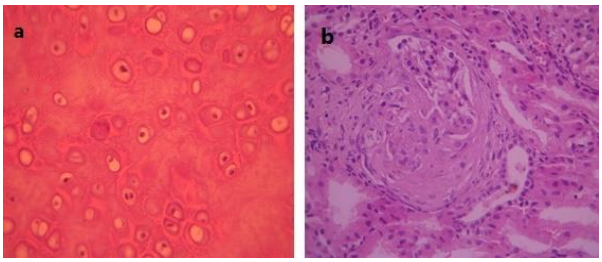

Figure 2. a) Trichrome staining of cartilaginous tissue; b) Hematoxylin and eosin stain (H&E) of kidney tissue

Our patient was managed with intravenous steroid pulse therapy (methylprednisolone of 1 g/day for 3 days), prednisolone, and cyclophosphamide. Her condition improved markedly as blood chemistries and estimates of glomerular filtration rate (GFR) demonstrated. Her 24-hour urine protein decreased to 620 mg (the previous was 1600 mg), and hemoglobin level that was 8.8 mg/dl increased to 14 mg/dl. She was discharged from hospital setting, and was put on regular follow-up program.

Discussion

RP is necrotizing episodic cartilage inflammation of unknown etiology with a spectrum of clinical features occupying different system and organs (2). An association between RP and HLA-DR6 and HLA-DR4 has been documented. There is no specific test for diagnosis of RP; thus the diagnosis is based on the multidisciplinary evaluation of the patient (5, 6).

The frequency of renal involvement in the

setting of RP is generally unknown. Chang-Miller et al. reported renal segmental necrotizing glomerulonephritis and mesangial expansion with cellular proliferation as the most common nephrology finding (3). Tubulointerstitial disease and IgA nephropathy are other renal lesions reported in RP; however, overall glomerulonephritis remains rare occurrence in the course of RP (7).

The laboratory evidence is neither conclusive nor specific. Although, clues of chronic anemia, episodes of leukocytosis thrombocytosis, and elevated levels of ESR and CRP has been detected; laboratory findings remain an inconclusive and occasionally incoherent component of disease (1).

RP is characterized by inflammation in cartilaginous structures as well as other anatomic locations including costal cartilage heart, eye, nose, airway, and kidney (1). The most common onset sign is external audit inflammation. In the absence of positive histological records, parameters of McAdams criteria is mandatory for establishment of RP diagnosis (2 major, or 1 major and 2 minor criteria), and in the presence of positive histological records, presence of one of features is enough to confirm the diagnosis (2).

McAdams criteria is divided in two groups, major and minor criteria. Major criteria are inflammatory episodes involving auricular cartilage, inflammatory episodes involving tracheobronchial cartilage, and inflammatory episodes involving nasal cartilage; minor criteria includes ocular inflammations (keratitis, conjunctivitis, episcleritis, or uveitis), hearing loss, vestibular dysfunction, and seronegative arthritis (8). The course of disease is preceded by unspecific constitutional signs including fatigue, malaise, and fever which may postpone RP diagnosis.

Based on McAdams criteria, the criteria of our patient were as inflammatory episodes involving auricular cartilage, conjunctivitis, and seronegative arthritis.

The most common presenting feature of RP is unilateral or bilateral auricular chondritis. Because of our patient manifestations, she underwent a complete nephrology workup. Frequent urine analysis was performed, and episodes of proteinuria and hematuria were detected. Urine culture was negative. Pathologic observation of renal biopsy containing five glomeruli revealed one total sclerosis, and two segmental sclerosis (9). Immunofluorescence microscopic study revealed anti-glomerular basement membrane IgM in 50% of observed tissue plus nonspecific and weak IgG and C3 deposition. These findings underlined classic variant of focal segmental glomerulosclerosis.

The etiology of RP is not fully understood. Electron microscopic studies and animal models have supported the role of degenerative enzymes in the process of cartilage destruction (1, 4). A dysregulation in autoimmune reaction to type 2, 9, and 11 collagen has been reported, too (5, 10). Furthermore, an over expression of HLA-DR4 antigen has been detected in a RP case (6). For treatment of these patients, some show good response to nonsteroidal anti-inflammatory drugs (NSAIDs), but some need corticosteroids and immunologic agents (9).

Renal involvement in the setting of RP is mainly important, and need close observation of these patients. Due to potential loss of renal function in focal segmental glomerulosclerosis in RP, and potential progress to end-stage renal disease (ESRD) in the course of focal segmental glomerulosclerosis, the nephrology workup in patients with hematuria or proteinuria along with RP is mandatory.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

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