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Granulomatous Polyangitis in a 17 Years Girl with Hemoptysis: A Rare Case Report

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Introduction

Granulomatosis polyangitis (GPA), formerly known as Wegner granulomatosis, is a rare multisystem autoimmune disease of unknown etiology. GPA is one of the ANCA-associated vasculitides and has a predilection for the upper and lower respiratory tracts and the kidneys. Although most cases of WG occur in adults with a peak age of presentation 64-75 years, it does develop in children, with a mean age at diagnosis of 14 yr[1]. In children, the estimated incidence is approximately 0.1:100,000[2]. There is a female predominance of 3-4: 1. Its hallmark features include necrotizing granulomatous inflammation and pauci-immune vasculitis in small- and medium-sized blood vessels.

Case Report

We present a case of a17yrs female adolescent, who presented with fever for 1 month and cough for 15 days. Fever was high grade, max 104.f daily spikes. There was history of productive cough associated with one episode of hemoptysis. On general physical examination, there were some hyper-pigmented lesions over both arms and legs. Rest of the systemic examination was normal. Hearing and eye evaluation was also normal. On investigations blood counts were normal but ESR was highly elevated upto 130mm/Ist hr, RFTs/LFTs/Electrolytes were normal. Urine shows hematuria with 250RBCs per microlitre. Chest xray showed multiple dilated cystic lesions in bilateral lung fields (as shown in figure 1). CT scan showed multiple cavitatory lesions in B/L lung fields with patches of consolidation in B/L upper lobes (figure 2-4).

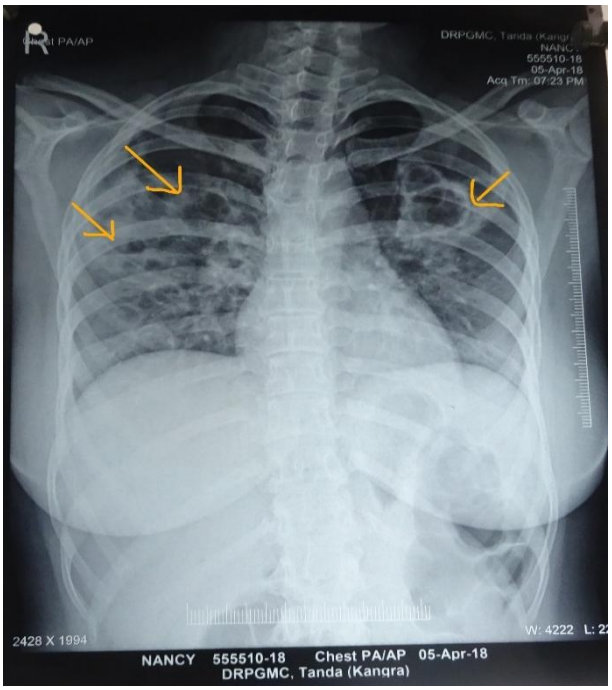

Figure 1: X ray image showing multiple cystic lesions in bilateral lung fields.

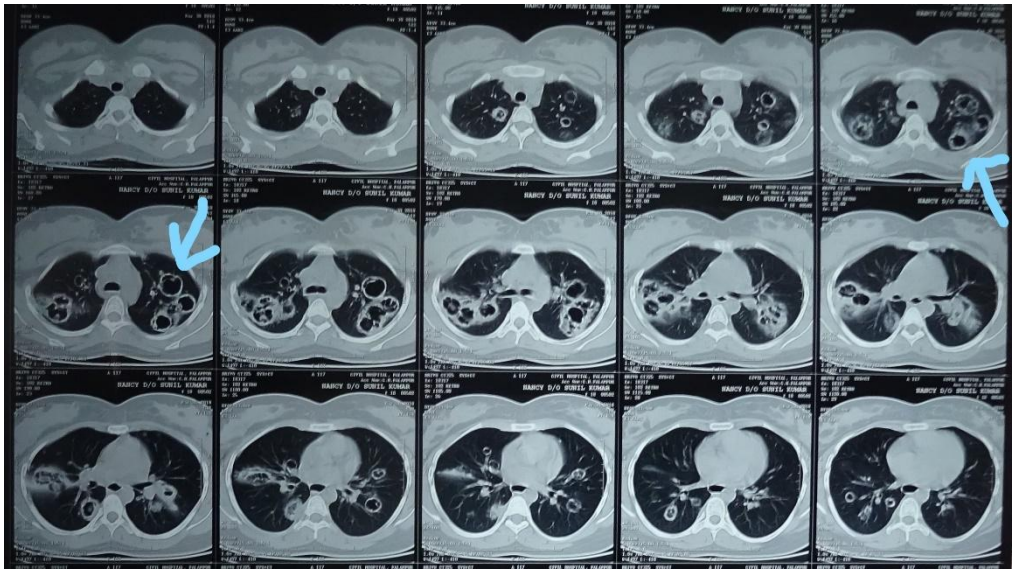

Figure 2: CT scan chest showing multiple nodulo-cystic lesions scattered over bilateral lung fields in axial view.

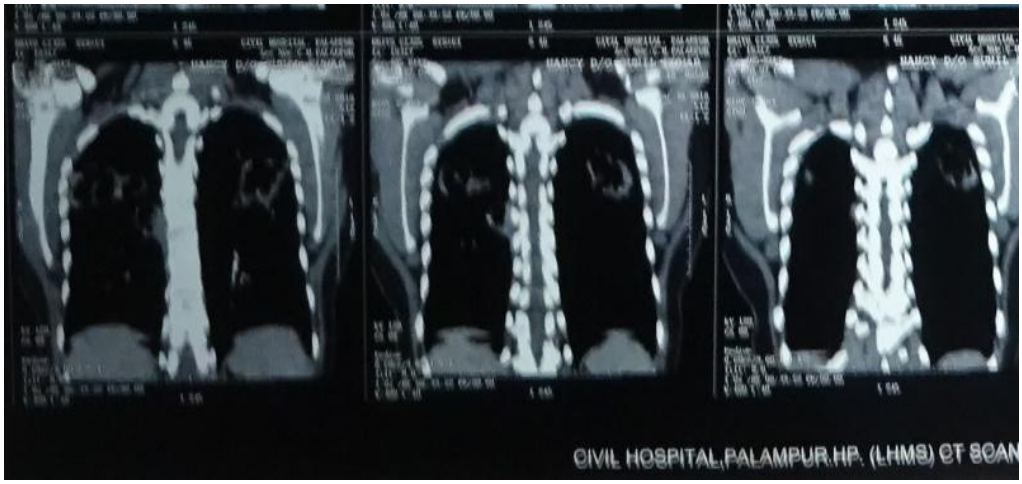

Figure 3: CT scan chest, anterior view of the same patient.

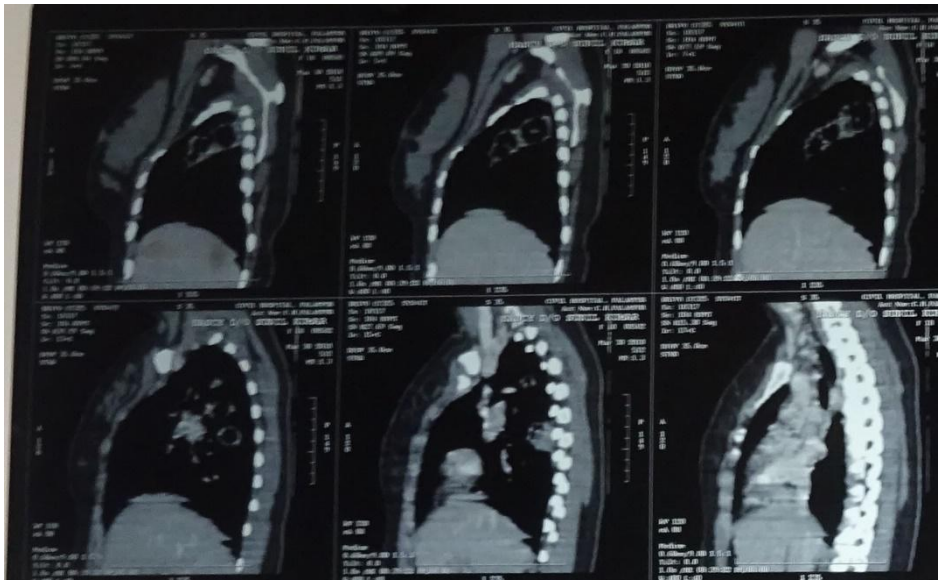

Figure 4: CT scan chest lateral view of the same patient showing multiple nodulocystic lesions.

Her Blood culture and senstivity, sputum for AFB were negative, cANCA was positive (1:40).

Course and management: Based on long duration fever with cough, hematuria and cANCA positivity, GPA was diagnosed according to EULAR/PRES criteria[3] and child was started on methyl prednisolone pulse therapy followed by cyclophosphamide pulse therapy along with leuprolide. Remission was achieved within two weeks and was maintained on oral prednisolone and monthly cyclophosphamide pulse therapy for next 6 months. Presently she is on low dose steroid maintenance therapy and has shown no signs of relapse for last 18 months.

Discussion

Severe, untreated GPA is associated with a very high (>90%) mortality rate hence early diagnosis and treatment is necessary for favorable outcome. Historically, patients with untreated GPA had a mean survival of 5 months from diagnosis; the mortality rate was 82% at 1 year. With the advent of cytotoxic therapy, patient survival in GPA markedly improved. With current treatments, the 5-year survival rate ranges from 74-79%[4]. Unfortunately, relapse is common in GPA. Typically, up to half of patients with GPA experience relapse within 5 years[5].

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