Problem 25th Kartik, 2059 (MD)

# An Eight-Year-Old Girl with Fever, Hemoptysis, and Pulmonary Consolidations

## Presentation of Case

An eight-year-old girl was admitted to the hospital because of fever, cough, hemoptysis, and pulmonary consolidations.

She had been well until six days before admission, when sore throat developed, with mild fever, a dry cough, and anorexia, and she began to vomit once daily. Three days before admission she began to cough up bright red blood and had right-sided otalgia and a sore throat; the low-grade fever persisted. The hemoptysis and dyspnea worsened, and she was taken to Bharatpur hospital, where chest radiographs showed pulmonary consolidations. She was treated with IV penicillin and gentamycin and the following day she was transferred to Kanti hospital.

The girl was an only child. Her immunizations were up to date as of EPI schedule. She had had two hospital admissions in the past for high fever diagnosed as typhoid and pneumonia.

During the eight months before admission, the girl had had an intermittent papular rash that involved her arms and legs and that had been called a "heat rash" by a physician; it disappeared for a month and then recurred at about the time of her current illness. When she was three years old, she was treated for urinary tract infections on two occasions. Five months before admission, the results of a voiding cystourethrographic study performed at TUTH to evaluate recurrent, painless hematuria were normal, and a renal ultrasonographic examination showed no abnormalities; the symptom was believed to be benign.There was no history of asthma, drug allergy, long-term use of medications, weight loss, or hematochezia. There was no family history of tuberculosis.

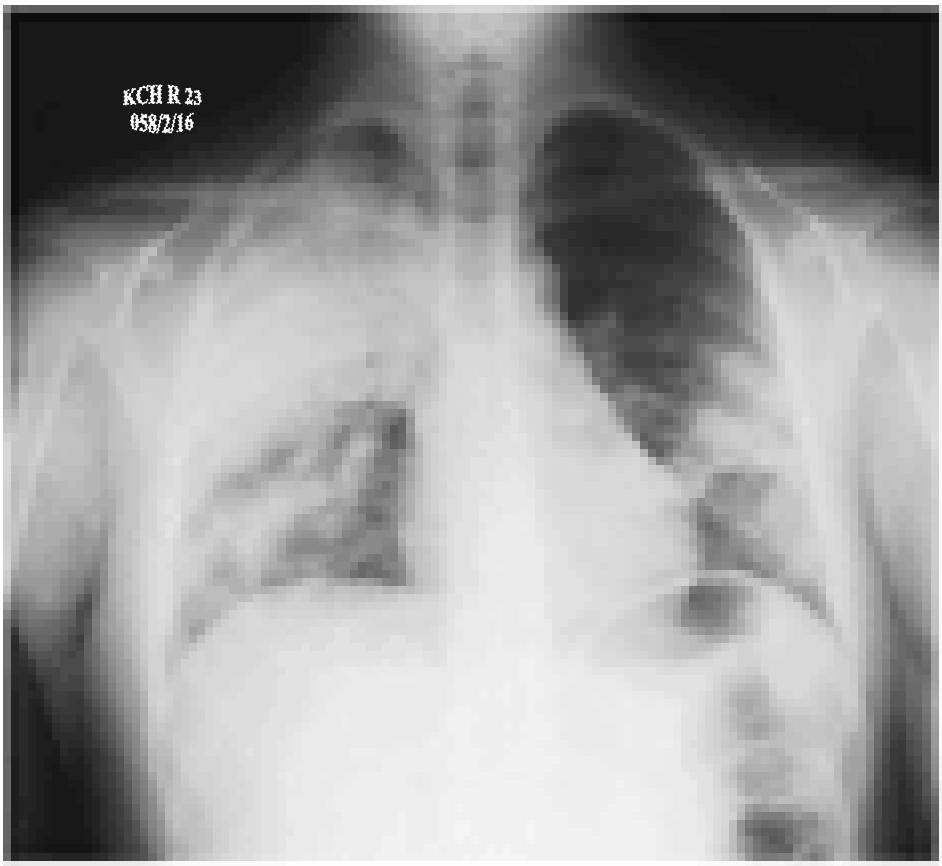
When arrived in the KCH: temperature was 38.3°C, the pulse was 142, and the respirations were 33. The blood pressure was 110/60 mm Hg. The body weight was 27 kg and the height 120 cms.

On physical examination, the girl was pale and panting and appeared acutely ill. A maculopapular rash that were brown, ovoid, and minimally elevated; for the most part the rash spared the face, hands, and feet. According to the girl's parents, each lesion had begun as a macule, that after several days had become a nonblanching papule, scabs then developed, without frank ulceration or vesiculation, and the lesions resolved. In addition, there was a circle, about 1 cm in diameter, of small vesicles, each 1 to 2 mm in diameter, on the medial aspect of the left thigh. No petechiae, telangiectases, or oropharyngeal lesions

were found, and there was no lymphadenopathy. There were decreased breath sounds over the right upper lobe, with bronchophony; no crackles or intercostal retractions were detected. The heart sounds and the abdomen were normal; a stool specimen was negative for occult blood. No sign of synovitis was evident. Neurologic examination revealed no abnormalities. ON ENT examination revealed pus in the right middle ear and a serous effusion in the left. The pharynx and tonsillar pillars were erythematous. Shotty posterior lymph nodes were palpated bilaterally. The larynx appeared normal.

In ER the investigations performed showed: TLC: 7.400/cmm; P:66; L: 29;M:4; E:1; Hb: 10.5G%;The urine was normal except that the sediment contained 5 to 10 white cells and 3 to 5 red cells per low-power field. Laboratory tests were performed subsequently showed ESR 116 mm /hour;Platelets: 4,10,000/cmm; Prothrombin time:14.6 sec (normal range is 11.1-13.1 seconds) The levels of conjugated and total bilirubin, electrolytes SGPT, SGOT, urea and creatinine were normal.

Chest radiographs showed complete opacification of the right upper lobe and confluent opacities in both lower lobes; no pleural effusion was evident, and the heart, mediastinum, and bony structures appeared normal.



Therapy with ceftriaxone was continued, and paracetamol were added to the regimen. On the evening of admission, the temperature rose to 38.9°C. The hemoptysis persisted, although the patient never coughed up sputum. On the second hospital day, the temperature rose to 38.1°C, and the respiratory rate ranged between 30 and 56 breaths per minute. Another specimen of urine, contained no protein or white cells, but there were many red cells, some of which were dysmorphic, and one red-cell cast. The results of laboratory tests are shown Preliminary examination of a blood culture, a urine culture, and a throat culture revealed no organisms. The findings on two sets of chest radiographs remained unchanged.

## Discussion

The most important aspects of the child's history and presentation are her chronic renal problems and acute pulmonary hemorrhage. This constellation of findings places the illness in the category of pulmonary–renal syndromes. Primary pulmonary–renal syndromes include Goodpasture's syndrome, systemic lupus erythematosus, and small-vessel vasculitides The chronic and recurrent skin findings and the problems affecting the ears are also striking. The abnormal urinary sediment five months before admission are evidence of long-standing, rather than acute, disease.

### Goodpasture's Syndrome

Goodpasture's syndrome is characterized by pulmonary hemorrhage and glomerulonephritis in patients with circulating antibodies against glomerular basement membrane or with linear immunofluorescent staining of IgG in the basement membranes of pulmonary or renal tissue. The prominent involvement of the skin, ears, and sinuses in this child, together with the markedly elevated erythrocyte sedimentation rate, strongly argues against a diagnosis of Goodpasture's syndrome.

### Systemic Lupus Erythematosus

Systemic lupus erythematosus has been well documented in children, especially young girls, and is commonly associated with rashes.

In the case under discussion, the underlying nephritis, the polymorphous rash and history of erythema multiforme, and the markedly elevated erythrocyte sedimentation rate are all consistent with a diagnosis of systemic lupus erythematosus. However, because the patient has no photosensitivity, no arthralgias, less severe renal dysfunction than would be expected in a person with systemic lupus erythematosus, and a rash that is atypical of the disease, systemic lupus erythematosus is an unlikely diagnosis.

Small-Vessel Vasculitides

The final group of diseases associated with pulmonary–renal syndromes are the small-vessel vasculitides. These include Wegener's granulomatosis, microscopic polyangiitis, the Churg–Strauss syndrome, Henoch–Schönlein purpura, and cryoglobulinemia. This child's clinical picture does not fit either Henoch–Schönlein purpura or cryoglobulinemia. The Churg–Strauss syndrome can cause the findings described in this case; however, the absence of peripheral-blood eosinophilia and of a history of asthma essentially rules out this diagnosis.

Both Wegener's granulomatosis and microscopic polyangiitis are pauci-immune vasculitides that involve the medium and small arteries, arterioles, and venules and capillaries and that do not involve substantial deposition of immune complexes in the tissue. The presence of granulomas in Wegener's granulomatosis distinguishes it from microscopic polyangiitis, and the expression of antineutrophil cytoplasmic antibodies (ANCA) differs in the two diseases. The clinical presentation of patients with these diseases, however, can be indistinguishable. In fact, cases have been reported in which patients who initially presented with microscopic polyangiitis without granulomas subsequently had granulomatous lesions and thus were later considered to have Wegener's granulomatosis.

Wegener's Granulomatosis

Criteria for the diagnosis of Wegener's granulomatosis include nasal or oral inflammation, abnormal findings on chest radiographs, abnormal urinary sediment, and granulomatous inflammation within an arterial wall or in a perivascular distribution. The upper respiratory problems associated with Wegener's granulomatosis include rhinorrhea, nasal mucosal ulceration or crusting, serous otitis media, and sinus pain or drainage.

Pulmonary disease develops in nearly three quarters of children with Wegener's granulomatosis, and cutaneous manifestations develop in approximately half at some point in the course of their illness. Cutaneous lesions, which may precede other systemic manifestations by a period of months, occur most commonly on the legs. The predominant skin lesion in small-vessel vasculitis syndromes is palpable purpura. Other lesions may be vesicular, papular, or nodular, and urticaria may be seen. Renal manifestations, including proteinuria and the

presence of red cells and red-cell casts in the urine, are seen in 60 to 80 percent of patients with Wegener's granulomatosis. At the time of presentation, the renal involvement is usually asymptomatic, but even in such cases, glomerulonephritis can progress rapidly.

Microscopic Polyangiitis

The clinical manifestations of microscopic polyangiitis include glomerulonephritis, weight loss, vasculitic rash, fever, arthralgias, mononeuritis multiplex, and pulmonary disease (including pulmonary hemorrhage). Both the upper and lower airways may be involved. Ear and sinus involvement is much less common in microscopic polyangiitis than it is in Wegener's granulomatosis.

Diagnostic Tests for Small-Vessel Vasculitides

Tissue Biopsy

Antinuclear Cytoplasmic Antibodies

Summary

In summary, this patient had a pulmonary–renal syndrome, and the probable diagnosis is Wegener's granulomatosis or microscopic polyangiitis. The diagnostic procedure consisted of serologic tests for anti–glomerular basement membrane antibodies, antinuclear antibodies, anti–double-stranded DNA antibodies, and ANCA (with specific testing for antibodies directed against proteinase 3 and myeloperoxidase

## Clinical Diagnoses

Necrotizing pneumonia with hemorrhage.

? Vasculitis (Wegener's granulomatosis or microscopic polyangiitis).