

10-YEAR-OLD GIRL WITH RECURRENT PNEUMONIA

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MEDICAL HISTORY OF PATIENT DI

- Previously healthy child, 1st pneumonia at age of 5 yrs
- Within following 5 yrs **9 episodes of pneumonia**
 - fever, WBC >20.000, ↑ CRP, chest auscultation findings
 - 1-2 episodes per year
 - **in-between** rare symptoms (cough/wheezing), CXR normal
- Mostly left lower lobe
- **Sputum cultures:** *M. catarrhalis*, *H. influenzae*, *S. aureus*, *C. albicans*
- **Chest Radiography:** bilateral infiltrations / consolidation
- **Treatment:** broad spectrum antibiotics for 14-21 days, response to therapy usually within few days

MEDICAL HISTORY

- **DOB:** 13/05/2000
- Female, term neonate, birth weight: 3600g
- No maternal illness, no neonatal problems, normal detachment of umbilical cord
- Breastfeeding for 1 year
- Immunization history: normal
- History of "bronchiolitis" at 6 months age
- No severe childhood diseases

FAMILY HISTORY

- Father: intermittent asthma
- Mother: healthy
- Older brother: allergic conjunctivitis/asthma
- Home environment: no smoke, no pets, no carpets, central heating

PHYSICAL EXAMINATION

- Growth charts: 50th percentile, maturational milestones in time
- Normal appearance, no signs of chronic disease (eczema, facial dysmorphies, pallor, no finger clubbing)
- S1, S2 normal, sinus rhythm
- No lymphadenopathy
- Normal cognitive status and neuromotor development

DIFFERENTIAL DIAGNOSIS

- **Anatomic abnormality:** eg, foreign body, bronchial compression by mediastinal adenopathy or vascular anomaly, bronchiectasis, bronchial cyst
- **Chronic Disease:** cystic fibrosis, immotile cilia syndrome, recurrent aspiration
- **Immunodeficiency**
 - **Secondary:** Underlying disease states like HIV/AIDS, diabetes mellitus, malignancy or nephrotic syndrome, immunosuppressive drugs, injury, previous surgical procedures, prematurity
 - **Primary:** Overall incidence 1/10,000: B cell, T cell, complement, phagocytic cell, combined

INVESTIGATION OF CHRONIC DISEASE

- Chloride test and DNA testing for cystic fibrosis: (-)
- Mantoux: always (-)
- HIV: (-)
- Normal total blood count, renal and hepatic function tests, urine tests

INVESTIGATION AT AGE 6,5 YEARS (4th episode)

- 1st Bronchoscopy:** Normal
- BAL: M. catarrhalis (R: ampicillin) + S. Aureus (R: penicillin), giant granulomatous phagocytes
- 1st HRCT:** normal
- Only finding: small left axillary lymphnode <8mm

- Investigation for sarcoidosis negative (ACE: within normal range, retinoscopy: normal, urine Ca/Cr: normal, no lymphadenopathy)

- Investigation for TB negative: TB culture: (-) Mantoux (-)

INVESTIGATION AT AGE OF 9 yrs (9th episode)

- 2nd Bronchoscopy:** Findings suggestive of chronic purulent bronchitis
- 2nd HRCT:** normal

BAL: *H. influenzae*, foam cells (DD: Sarcoidosis?)

Air tract lumen full of mucopurulent secretions, swollen mucosa, slight bronchomalacia of a small number of the segmental bronchi, mainly on the left, sometimes causing the bronchi to collapse during bronchoscopy.

IMMUNOLOGICAL TESTING

	2008	2009
IgG mg/dl	993	1220
IgG1	530	610
IgG2	198	233
IgG3	45	48
IgG4	11↓	23
IgA	128	146
IgM	102	94
IgE IU/ml	653↑	1453↑

Prick and Rast tests: negative

IMMUNOLOGICAL TESTING

- Complement factors C3, C4: within normal range
- Antibody response to vaccines: within satisfying levels for her age and previous vaccines

Lymphocyte immunophenotype

	%	Cells/ul
CD3+	68,6	1898
CD3+CD4+	66	
CD3+CD8+	28,8	
CD4+/CD8+	1,8	
CD19+	19	532
CD20+	16	
CD3+CD16+	9,2	
CD3+HLADR+ (on CD3+)	7,7	
CD3+CD16+(on CD3+)	0,68	

WBC:5320
LY: 52 %
Normal percentage and absolute count of T,B,NK cells
Normal CD4/CD8 ratio

TREATMENT SCHEDULE

- Under chemoprophylaxis with amoxicilline + clavulanic acid since 12/2009
- Free of pneumonias ever since

IS THIS AN IMMUNODEFICIENCY?

Thank you

