



CASE REPORT

SOFIA TANTOU

Specific Center & Referral Center for
Primary Immunodeficiencies – Paediatric
Immunology
"Aghia Sophia" Children's Hospital, Athens
Head Director : M.G. Kanariou

PATIENT PZ

- ✘ Male 6 ½ years old (dob: 2/12/1999) is being referred due to frequent infections and asthma
- ✘ Perinatal history : 5th child , 2nd child of a tween gestation
GA :33w , BW :1410 gr , Apgar score: 1⁰,5⁷
- Hospitalization in NICU due to :
 - ✘ Perinatal asphyxia
 - ✘ Prolonged rupture of membranes
 - ✘ Neonatal jaundice
 - ✘ Apnea
 - ✘ Anaemia
- Family history : negative

MEDICAL HISTORY

Hospitalization	Clinical condition	Laboratory findings	Diagnosis <i>Recommendation</i>
2000	Respiratory distress	Chest X-ray : bilateral hyperventilation	Bronchiolitis
2002	Fever cough	Chest X-ray : Consolidation of left inferior lobe	Pneumonia <i>1.Repeat X-ray</i> <i>2.Department of Allergy</i>
2003	1st Respiratory distress cough	Chest X-ray : Bilateral infiltration mainly at the left side Immunological findings : IgG:795 mg/dl IgA:255mg/dl,IgE:93 IU/ml	Asthma attack
	2nd Fever cough	Chest X-ray : Consolidation of left lower lobe Mycoplasma Abs : (+) Mantoux : (-)	Mycoplasma consolidation <i>Department of Allergy</i>

Hospitalization	Clinical condition	Laboratory findings	Diagnosis <i>Recommendation</i>
2003	3rd Fever cough	Chest X-ray : Retrocardiac infiltration Sweat test : (-)	Mycoplasma pneumonia
2005	Fever Cough Respiratory distress	Chest X-ray : Viral disease with atelectasis Mantoux:(-)	Bronchopneumonia
2006	fever	Immunological findings: IgG:555,IgM:76,8 IgA:283,IgE:56,5	Pneumonia Asthma <i>Bronchoscopy</i>
2007	Fever Abdominal pain Rash	Sputum culture: (+) Moraxella catarrhalis	Respiratory infection <i>Cellular immunity control</i>
2009	Sputum culture: (+) Candida		Respiratory infection on the ground of bronchiectasis

Hospitalization	Clinical condition	Laboratory findings	Diagnosis <i>Recommendation</i>
2003		Mucopurulent secretion-edematous mucosa easily bleeding, stenosis of right middle lobe-ligula opening Findings compatible with chronic purulent bronchitis BAL culture : H.influenza-candida albicans <i>Spirometry:</i> 1st)Mixed type pneumonopathy with prominent the obstructive element.	Pneumonia
2005		2nd)Incompatible with obstructive pulmonary disease <i>Chest CT:</i> Heterogeneous density of pulmonary parenchyma in middle and inferior pulmonary fields.Findings compatible with small airways disease.	Pneumonia
2006	fever	Immunological findings: IgG:555,IgM:76,8	Pneumonia Asthma <i>Bronchoscopy</i>
2007	Fever Abdominal pain Rash	Immunological findings : IgG:692,IgM:79,5,IgA:248,IgE:41,2 C3:111,C4:21,7 <i>Bronchoscopy:</i> Mucopurulent secretion-edematous mucosa, stenosis of right middle lobe-ligula opening BAL culture: Moraxella catarrhalis	Respiratory infection <i>Cellular immunity control</i>
2009	Sputum culture: (+) Candida		Respiratory infection on the ground of bronchiectasis

IMMUNOLOGICAL CONTROL (I)

	2003	2006	2007	2009	2010
IgG mg/dl	795	555 ↓	757	750 ↓	960
IgG1			498	480 ↓	533
IgG2			147	181	359
IgG3			37	37	38
IgG4			35	55	64
IgA	255	283	210	199	225
IgM	-	76,8	82	78	113
IgE IU/ml	93	56,5	27	31,7	

IMMUNOLOGICAL CONTROL (II)

	%	Cells/µl
CD3+	74,3	1986
CD3+CD4+	27,5	735
CD3+CD8+	39,7	1059
CD4+/CD8+	0,69	
CD19+	11,6	309
CD20+	11,5	302
CD3-CD16+	11,6	309
CD3+HLADR+ (on CD3+)	11	230
CD3+CD16+	2,2	58

WBC:6.360
LY:42 % NE:45 %MO:6 %
Normal percentage and absolute count of T,B,NK cells
Reversed CD4/CD8 ratio

IMMUNOLOGICAL CONTROL (III)

- ✗ Normal chemotaxis - phagocytosis
- ✗ Normal antibody response to vaccines
- ✗ Complement parameters : C3:105 mg/dl
C4: 33 mg/dl

OUR PATIENT PRESENTS...

- Normal Igs and IgG s/c concentrations (marginal low IgG)
- Normal numbers B,T NK
- Normal chemotaxis-phagocytosis
- Normal antibody response



Diagnosis

- Functional antibody deficiency with normal Igs concentrations ?
-?

PATIENT FOLLOW-UP

- ✗ 2009 : Immunoglobulin replacement therapy (subcutaneously)
- ✗ Free of respiratory tract infectious diseases



DISCUSSION

THANK YOU FOR YOUR TIME...

