

# **Lymphoma, vasculitis and arthritis in NBS patient**

**case report**

**Clinical Immunology Department,  
Russian Children Clinical Hospital**

- ✓ **R.P., born in 1991**
- ✓ **One child in the family**
- ✓ **No family history**
- ✓ **Microcephalia diagnosed after birth**
- ✓ **Dismorphic symptoms: receding forehead, prominent midface.**
- ✓ **Cafeau-lait-spots.**



# R.P. Lymphoma

- ✓ No serious diseases till 12 years 3 month (Sep. 2003).
- ✓ Sep. 2003 enlargement of all groups of Peripheral lymph nodes, fever 37.8-38.2° C. Antibiotics without effect.
- ✓ Admitted in Russian Clinical Children Hospital.
- ✓ NBS syndrome diagnosed: 657-del5 mutation.

# R.P. Lymphoma

- ✓ **At the time of admission hospital:**
  - **peripheral lymph nodes 8-10-12 mm**
  - **hepar + 3 cm. below rib arch,**
  - **spleen +3 cm below rib arch.**
  - **Blood count leucopenia –  $3.8-1.4 \times 10^9/l$ ,**  
**neutropenia**
  - **increased LDH,**
  - **IgA <6.67 mg/dl, IgG-325 mg/dl,**  
**IgM-39.8 mg/dl.**
  - **7.2% blast cells in bone marrow.**

# R.P. Lymphoma

- ✓ **Lymph node biopsy - Hodgkin disease (two labs)**
- ✓ **17-30.10.2003 – DAL-HD-90 protocol: Pred 60мг/м<sup>2</sup>, Vcr1,5мг/м<sup>2</sup> №1, Adr40мг/м<sup>2</sup> №1, Vp-16 125мг/м<sup>2</sup>м №4.**
- **Adverse events: 30.10 – mucositis, hypertemia, vincristine (oncovin) neuropatia**
- **Treatment: cephtriaxone, vanco, metronidazole, amycacine, G-CSF, ampho-B, IVIG.**

# R.P. Lymphoma

- ✓ **Lymph node biopsy** *diffuse large B-cell lymphoma, variant T-cell rich 3 group of risk, IV st (cervical, axillar, mediastinal, retroperitoneal lymph nodes and bone marrow involvement) the third lab (prof Dr.Parwadesh, prof Dr. Wacker Germany): All blast cells express CD20, reactive surrounding is CD3 positive. Blast cells are Ki-A10 negative.*

# R.P. Lymphoma

✓ **12 - 16.11.2003- AA(1):** MTX 5 g/ m<sup>2</sup> , Dexa 10 mg/ m<sup>2</sup> №5, VP 100 mg/ m<sup>2</sup> №2, ARA-C 150 mg/ m<sup>2</sup> №2, IFO 400 mg/ m<sup>2</sup> №5, Vcr 1,5 mg/ m<sup>2</sup> №1.

✓ **Adverse events:**

delayed MTX clearance (0.24 on 66 hours).

day +7 – neutropenic enterocolitis  
(Enterococcus, Staph),

intestinal hemorrhage, pancreatitis

✓ **Treatment:** cefepim+vanco+amicacine,  
Ampho-B, E-ACA, G-CSF.

# R.P. Lymphoma

- ✓ **28.11 - 02.12.2003 BB(2):** Dexa 10 mg/ m<sup>2</sup> №5, MTX 5 g/ m<sup>2</sup> , Adr 25 mg/ m<sup>2</sup> №2, chp 100 mg/ m<sup>2</sup> №5, Vcr 1,5 mg/ m<sup>2</sup> №1 . Concomitant therapy – cthephepim+fluconazole. Leikovorin.
- ✓ **Adverse events:**
  - delayed MTX clearance (4,68 on 42 hours)
  - Day +7 diarrhea, arterial hypertension.
  - **Therapy:** cehephepim+vanco+amicacin+AmphoB, GCSF, amlodipin+enalapril.
  - Day +9 wormiting, bloody diarrhea, oedema, arterial hypertension, strabismus, convulsions.
  - **Therapy** – carbopenem +vanco +amycacin +AmphoB +metronidazole, leikovorin, G-CSF, dobutamin, nifedipin.
- ✓ **28.11 – clinical and laboratory remission. 1% blast cells in bone marrow**



# R.P. Lymphoma

- ✓ **19.12.03 - 23.12.03 CC(3):** Dexa 10 mg/ m<sup>2</sup> №5, Vcr 1,5 mg/ m<sup>2</sup> №1, VP 100 mg/ m<sup>2</sup> №3, cytozar high dose - 4 g/ m<sup>2</sup> №2. Concomitant therapy: cephperason/sulbactam +fluconazol.
- ✓ **Adverse events:**
  - **Day +6** arterial hypertension, sepsis, neutropenic enterocolitis (*Candida non-albicans*), mucositis.
  - **Therapy:** cehephepim+vanco+amicacin+AmphoB, GCSF, amlodipin+enalapril.
  - **Day +9** wormiting, bloody diarrhea, oedema, arterial hypertension, strabismus, convulsions.
  - **Therapy** – carbopenem +vanco +amycacin +AmphoB +metronidazole, leikovorin, G-CSF, dobutamin, nifedipin.

# R.P. Lymphoma

- ✓ **27.01.04 - 31.01.04. OP (4) (Vcr 1.5 мг/м<sup>2</sup> +Pred 40 мг/м<sup>2</sup> ) + Mabthera 500 мг/м<sup>2</sup> (4) + ITLP №2 (MTX+Dexa+ARA-C).**  
cephepim+vanco+amikacin+valcyclovir,  
nifedipin, enalapril.
- ✓ **Adverse events:**
  - **Febrile neutropenia, arterial hypertension. Day +14 Herpes Zoster.**
- **Therapy: cepepim+vanco+amicacin+valcyclovir, nifedipin, enalapril.**

# R.P. Lymphoma

- ✓ 24.02.04 - 01.03.04 COP (5): (Cph 300мг/м<sup>2</sup>+ Vcr 1.5 мг/м<sup>2</sup> +Pred 40 мг/м<sup>2</sup> ) +Mabthera 500мг/м<sup>2</sup> + ITLP (MTX+Dexa+Cytozar).
- ✓ 09.03.04 Mabthera 500мг/м<sup>2</sup>
- ✓ 19.03.04 - 25.03.04COP(6) + Mabthera 500мг/м<sup>2</sup> + ITLP №2 (MTX+Dexa+Cytozar).

Without adverse reactions.

# R.P. Vasculitis+Arthritis

- ✓ **Since April 2004 -- erythematouse rash (spots with proliferative component).**
- ✓ **Skin biopsy showed vaskulitis.**
- ✓ **Local treatment with prednisone without effect.**

# R.P. Vasculitis+Arthritis

- ✓ **October 2004** developed polyarthritis. Treatment – prednisone 2 mg/kg with good effect for arthritis and vaskulitis. The dose of prednisone gradually decreased till 5 mg/day.





