Cystic Leukoencephalopathy as a manifestation of Congenital Rubella Syndrome

Manjinder S Randhawa¹, Sumeet Dhawan¹, Renu Suthar¹, Sameer Vyas², Mini P Singh³, Naveen Sankhyan¹

¹Department of Pediatrics, ²Department of Radiodiagnosis, ³Department of Virology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

Introduction

The neurological manifestations of Congenital Rubella Syndrome (CRS) include;

- psychomotor retardation,
- chronic meningoencephalitis, parenchymal necrosis and vasculitis with calcifications.

Cystic leukoencephalopathy has rarely been reported with CRS.

We present a series of 3 cases with cystic leukoencephalopathy as a manifestation of CRS.

Case definitions

All cases fulfilled the following criteria:

- Rubella IgM positive
- CMV IgG/IgG negative
- Toxoplasma IgG/IgM negative
- HSV IgG/IgM negative
- HIV ELISA negative
- Maternal Rubella IgG positive

References

Sawhani V, et al. Magnetic resonance imaging findings in a case of congenital rubella encephalitis. Can J Infect Dis Med Microbiol. 2013;24:e122–3.

Uggetti C, et al. Aicardi-Goutières Syndrome: Neuroradiologic Findings and Follow-Up. Am J Neuroradiol. 2009 1;30:1971.

Case 1

- Presented at 3 months of age
- Dystonias - Abnormal involuntary posturing - 1½ Months
- Bilateral sensory-neural hearing loss.
- No cataract.
- No structural cardiac defect.
- Microcephaly and retrognathia.

Case 2

- Presented at 6 weeks of age
- Congenital cataract.
- 4 mm non-restrictive PDA with severe PAH
- Hepatomegaly and microcephaly.
- Skull - severe over-riding of sutures
- No hearing loss.

Case 3

- Presented at 2 Months
- Severe microcephaly,
- Patent Ductus Arteriosus,
- Delayed attainment of milestones and sutural over-riding
- Bilateral sensory-neural hearing loss.
- No cataract.

Neuroimaging Characteristics

	Case 1	Case 2	Case 3
Calcifications	Present, Periventricular	Present, Periventricular and basal ganglia	None
White matter changes	Cystic changes, Fronto-parieto-temporal	Hyperintense on T2/FLAIR, Periventricular, Cystic	Cystic changes, parieto-occipital
Temporal white matter changes	Cystic changes	Cystic changes	None
Temporal cysts	Present	Present	None

Conclusions

- Cystic leukomalacia and calcifications are rare manifestations of Congenital Rubella Syndrome
- **Leukoencephalopathy with temporal cysts in CRS mimics ; Aicardi-Goutieres Syndrome and Congenital CMV infection**
- Antenatal history of rash, fever or arthralgia is not found in all cases