P504 SANJAD-SAKATI-RICHARDSON-KIRK SYNDROME

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Early diagnosis of rare genetic disorders plays a vital role in preventing the disorder through appropriate genetic counseling. Sometimes early recognition of a rare disorder can help in reducing the associated morbidity and mortality.

The syndrome of severe growth retardation, mental retardation, and chronic hypocalcemia caused by hypoparathyroidism was first reported by Sanjad, Sakati, and Abu-Osba in 1988.

Sanjad, Sakati, and Abu-Osba presented a part of a full description of the syndrome in five infants at the 58th Annual Meeting of the Society for Pediatric Research, Washington DC, May 1988. Later, they published a more completed description of the syndrome in 1991. The paper of 1991 which was authored by more authors, and included twelve patients.

However, the syndrome was first fully described in 1990 by Ricky J Richardson from the Sick Children Hospital of Great Ormond Street in London, and Jeremy MW Kirk from St Bartholomew's Hospital in London.

Richardson and Kirk emphasized that this association of a previously undescribed congenital anomalies represented a new syndrome that was observed in eight children of middle eastern origin.

They thought that early recognition of this rare disorder may decrease the associated morbidity and mortality.

The total number of the reported patients with Sanjad-Sakati-Richardson-Kirk syndrome is 103.

Sanjad-Sakati-Richardson-Kirk syndrome has not been reported before in Iraq. The main aim of this paper is to report the first case of this syndrome in Iraq which is the case number 104 in the world.

Before referral, the child didn't receive management that can be described as appropriate.

The hypocalcemic seizure, he had was treated with low dose vitamin D followed with low dose of one alpha hydroxycholecalciferol (2 drops daily), and the boy continued to have seizures.

The calcium level remained below 7 mg/dL, but it was ignored by the treating physicians, and electroencephalography was performed and showed frequent focal epileptic discharges more prominent in central leads and mild slowing of cerebral activity.

The child was treated with anticonvulsant medications mostly sodium valproate which reduced the seizures, but didn't stop.

After referral, the child's hypocalcemia was treated with the appropriate doses of one alpha hydroxycholecalciferol. Calcium level was maintained above 8 mg/dL. Seizures stopped and sodium valproate was stopped.

P505 BRAIN ABSCESS AS A FIRST MANIFESTATION OF ATRIAL SEPTAL DEFECT

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A 6-year-old boy was admitted to our hospital due to a routine cardiac surgery of primum atrial septal defect (ASD) with cleft mitral valve. His medical history started seven months before, after he was admitted into the local hospital because of fever with chills, headache, nausea, stiff neck and vomiting. Diagnosis of meningitis was made, and antimicrobial treatment was initiated. Five days after, left side hemiparesis was detected. MSCT was performed and showed multiple brain abscesses on right part of basal ganglia and on the left along the temporal horn of ventricle system with a "mass effect". Furthermore, the threatening brain herniation was noticed. On that same day, he was transferred into our hospital for brain abscess drainage. Echocardiography was performed because of a still heart murmur and the diagnosis of primum ASD with cleft mitral valve was discovered. An indication for operative cardiac surgery after neurosurgical treatment was set. Control brain MR was been performed two weeks. On imaging there was no improvement, but muscular strength and general condition were better and the second drainage of brain abscesses with stereotactic method was performed. Streptococcus intermedius and mixed anaerobes were isolated from intraoperative smear. Antimicrobial treatment was in total of 6 weeks. Postoperative MSCT scan showed a significant regression of brain abscesses. Dental treatment for caries was done.

Six months later he went through an open heart surgery where an ASD primum was closed with pericardial patch and mitral valve plastic was done. Postoperative period was uneventful.

Conclusion Brain abscesses are rare, but serious, life-threatening neurological entities. The abscesses are usually deep seated and thin walled. Right parietal lobe is often more affected because of the direct blood flow to this area. Acyanotic congenital heart diseases are rare cause of brain abscess in children; however, an ECHO should be done as routine examination to exclude one.

P506 THE PATTERN OF PRESENTATION IN CHILDREN WITH HERPES SIMPLEX ENCEPHALITIS¹Jennifer Cox*, ¹Elizabeth Murphy, ¹Laura Whitla, ^{1,2}David Coghlan, ^{1,2}Montaseur Nadeem. ¹Tallaght University Hospital, Dublin, Ireland; ²Trinity College, Dublin, Ireland

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Introduction HSE is estimated to occur in approximately 1 in 250,000 to 1 in 500,000 individuals per year, with approximately one third of cases occurring in children and adolescents. Poorer prognosis and neurological sequelae are associated with delayed treatment of HSE.

We retrospectively reviewed all cases of HSV meningoencephalitis admitted to our centre over a ten year period from Jan 2008 to Jan 2018. Four cases were identified based on PCR confirmation of HSV 1 or 2 in cerebrospinal fluid samples. Here we discuss the clinical presentation, as well as electroencephalogram, neuroimaging and clinical outcomes, to highlight the importance of early recognition and timely treatment of such cases.

Case series Case 1: Nineteen month old male infant presented with prolonged focal febrile seizure, on a preceding history of pyrexia and lethargy for one day, and cough for one week. EEG showed excess slowing over both posterior regions, more marked on the left posterior region, consistent with bilateral cerebral dysfunction without epileptiform discharges. MRI