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Case Report

Cerebellitis and Concomitant Acute Hydrocephalus in a Child: Case Report

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Summary

Acute cerebellitis is a severe neurological disease characterized by mild or high-grade fever, nystagmus, tremor, truncal ataxia, dysarthria, headache, and altered mental state. The diagnose can be established on the basis of clinical symptoms, radiological imaging and laboratory findings. Acute life threatening hydrocephalus can be seen as a complication of acute cerebellitis associated with obstruction at the level of the fourth ventricle. Neurosurgical procedures as a life-saving intervention can be required which range from external ventricular drainage to ventricular peritoneal shunt and posterior fossa decompression. In this report we present a case of cerebellitis and concomitant acute hydrocephalus in a child which was treated succesfully with external ventricular drainage and medical treatment and also discuss the main features of this rare but important concomitance.

Key words: Cerebellit, Obstructive, Hydrocephalus, Child

Pediatrik Bir Hastada Serebellit ve Akut Hidrosefali Birlikteliği : Olgu Sunumu

Özet

Akut serebellit orta veya yüksek ateş, nistagmus, tremor, trunkal ataksi, dizartri, başağrısı ve bilinç değişikliği ile karakterize ciddi bir nörolojik hastalıktır. Tanı klinik semptomlar, radyolojik görüntüleme ve laboratuvar bulguları ile birlikte konur. Dördüncü ventrikülün obstrüksiyonu ile birlikte görülen akut serebellitlerde ani hayatı tehdit eden hidrosefali komplikasyonu görülebilir. Eksternal ventriküler şant uygulaması,ventrikülöperitoneal şantlama ve posterior fossa dekompresyonu gibi hayat kurtarıcı nöroşirürjikal müdahaleler gerekebilir.Bu yazıda başarılı bir şekilde eksternal ventriküler drenaj ve medikal tedavi uygulanan serebellit ve akut hidrosefalinin birlikte görüldüğü pediatrik bir olguyu sunduk ve bu ender görülen ancak önemli durumu ana hatları ile tartıştık.

Anahtar Kelimeler: Serebellit, Obstrüktif, Hidrosefali, Çocuk

INTRODUCTION

Acute cerebellitis is a severe neurological disease characterized by mild or high-grade fever, nystagmus, tremor, truncal ataxia, dysarthria, headache, and altered mental state[(5)]. The diagnose can be established on the basis of clinical symptoms, radiological imaging and laboratory findings. Acute cerebellitis can, rarely, cause obstructive hydrocephalus with compression of the fourth ventricle

and obliteration of the basal cisterns[9]. Early diagnose and urgent surgical intervention if necessary can be life saving in this hazardous disease. In this report we present a 6-year-old boy with cerebellitis and acute concomitant hydrocephalus and discuss the main features of this rare but potentially fatal concomitance.

CASE PRESENTATION

A previously healthy 6-year-old boy presented with fever, headache, and vomiting 4 days after a brief upper respiratory tract illness. On admission, he was intermittently lethargic and irritable, and complained of continuous nausea and headache. His neck was supple, and a fundoscopic examination revealed bilateral papilla edema. There was mild right-sided dysdiadochokinesia and scanning speech. No gait testing was done because of the child's reluctance to move. Computed tomography (CT) scanning on admission revealedlow-density areas in the bilateral cerebellar hemisphere. There was obstructive hydrocephalus with compression of the fourth ventricle and obliteration of the basal cisterns (Fig. 1-2). Magnetic resonance (MR) imaging revealed increased signal on T2-weighted images in both cerebellar hemispheres, with associated edema and obstructive hydrocephalus. There was no enhancement following the administration of gadolinium (Fig. 3-4-5). Laboratory findings at the time of admission included leukocytosis, with a white cell count of 17.400 /mm3 and a C-reactive protein (CRP) level of 3,44 mg/dl. We diagnosed acute cerebellitis on the basis of the history of infection and the neurological findings. The patient was empirically treated with vancomycin, ceftriaxone, and acyclovir. We concluded that the boy's symptoms originated from obstructive hydrocephalus and therefore an external ventricular drainage(EVD) catheter was initially placed at the anterior horn of the right lateral ventricle 6 h after admission. Urgent posterior fossa decompression was also planned to perform if the symptoms persist. The cerebrospinal fluid (CSF) pressure was over 20 cmH2O, and laboratory examination of CSF revealed a protein level of 7,2 mg/dl, a glucose level of 100 mg/dl, and 5 white cells/mm3. All cultures, including CSF, urine and throat swab, were negative for fungi and bacteria. The blood culture, meningococcal and pneumococcal PCR and viral serology (CMV, EBV, VZV, HHV-6, HSV 2) were negative. Herpes Simplex Viruse (HSV) type 1 IgM antibody was positive in blood but not in CSF. Multiplex PCR CSF analyze for all viruses was negative . The patient's headache and vomiting resolved promptly with ventricular drainage. Treatment with intravenous pulse injection of methylprednisolone (1 g/day for 3 days followed by dose tapering) was also started and cerebellar signs gradually improved over the next 7 days. The EVD was clamped on hospital day 7 and removed on hospital day 10. By hospital day 13, his neurological examination had returned to normal with minimal ataxia and he was discharged to home.

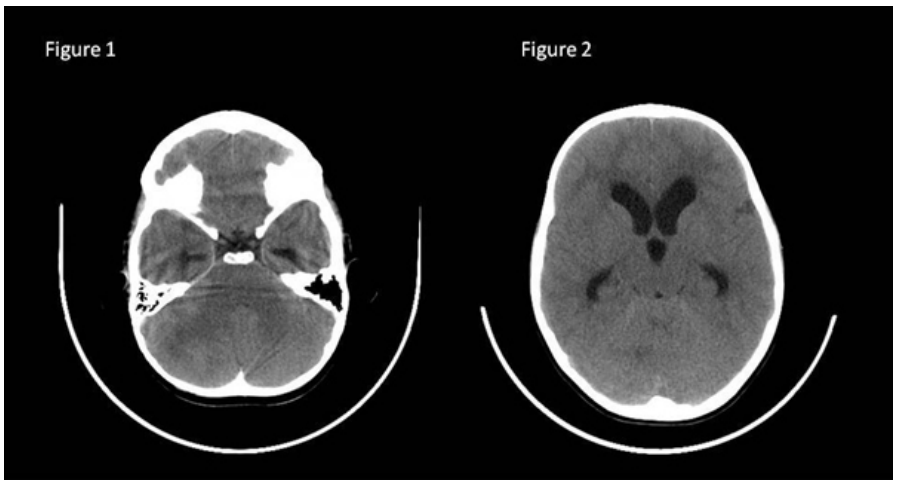

Figure 1-2: *CT scans on admission showing low-density areas in the bilateral cerebellar hemispheres and findings of obstructive hydrocephalus.*

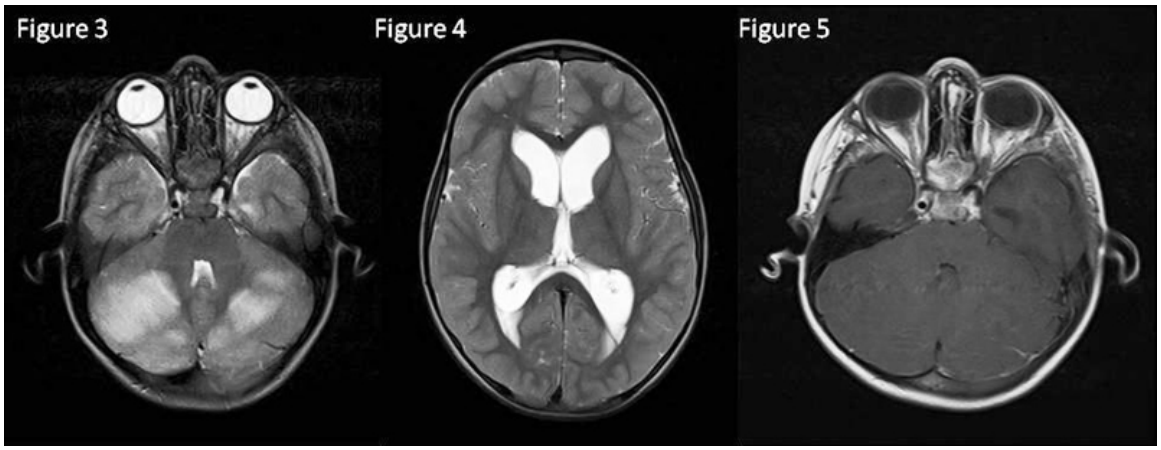

Figure *3-4-5: MRI at presentation: T2-weighted axial and post contrast T1-weighted images demonstrating high signal changes in both cerebellar hemispheres, modarate cerebellar swelling with moderate dilatation of the lateral ventricles without contrast enhancement.*

DISCUSSION

The diagnose of acute cerebellitis is based on clinical features along with neuroimaging findings[2,8]. The common symptoms are headache, vomiting and disturbances of consciousness as seen in our case. Sometimes fever, meningeal signs and occasionally brainstem signs are present. Ataxia may not always be a presenting feature[2,8]. Association with a variety of infectious agents has been described previously, including varicella zoster virus, Epstein- Barr virus, Bordetella pertussis, mumps virus, rubella virus, enteroviruses, Borrelia burgdorferi, Coxiella bernetii and M. pneumoniae[2,8,9]. But not rarely the agent organism remains unknown inspite of sophisticated serological tests as presented case[2,8]. Direct invasion and replication, an immune mediated post-infectious mechanism, cytokines release in the CNS and an autoimmune pathogenesis has been indicted as pathogenetic factors by previous studies[2,8,10]). The clinical course may be rapid or protracted. There does not appear to be a correlation between the organism implicated and the clinical

evolution[2]. Acute life threatening hydrocephalus has been previously reported as a complication of acute cerebellitis associated with obstruction at the level of the fourth ventricle[1,3,4,6,7,9]. It has been attributed to either compression of the fourth ventricle or progressive inflammatory subarachnoid space obstruction preventing CSF reabsorption or a combination of both these phenomena[1,3]. Fatalities have been reported in cases resulting from severe cerebellar swelling, with evidence of upward transtentorial and downward tonsillar herniation resulting in brainstem compression[3,7]. Neurosurgical procedures as a life-saving intervention can be required which range from external ventricular drainage to ventricular peritoneal shunt and posterior fossa decompression[3,4]. In the presence of obstructive hydrocephalus there is a risk of upward herniation with ventricular drainage if the drain is set at too low pressures and if the patient is not monitored carefully. When the hydrocephalus occur secondary to progressive inflammatory subarachnoid space obstruction the posterior fossa decompression should be considered[1]. In our case, a rapid clinical response followed emergency ventricular drainage and posterior fossa decompression is not required. Althought there has been no consensus as to whether steroids should be given or not[2,4] pulse treatment with high-dose corticosteroids was effective in our case. Resolution of clinical symptoms and MR imaging changes followed the initiation of corticosteroid treatment. Early accurate diagnosis, close monitoring and urgent surgical intervention if necessary is very important in ensuring appropriate treatment of this potentially fatal disease[4]. Emergency treatment, including surgical management and the initiation of high-dose of corticosteroids without delay, resulted in a good outcome in our case. We recommend close monitoring of cerebrospinal fluid pressure monitoring for avoiding important risks such as upward transtentorial and downward tonsillar herniation.

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REFERENCES

1. *De Ribaupierre S, Meagher-Villemure K, Villemure JG, Cotting J, Jeannet PY, Porchet F, Roulet E, Bloch J. The role of posterior fossa decompression in acute cerebellitis Child's Nerv Syst 2005; 21: 970–974*
2. *Gayatri N, Tyagi A, Mahadevan U. Acute hydrocephalus in a child with Mycoplasma cerebellitis Brain Dev 2009; 31(8): 618-21*
3. *Hacohen Y, Niotakis G, Aujla A, Siddiqui A, McCormick D, Bassi S, Clarke A, Lim M. Acute life threatening cerebellitis presenting with no apparent cerebellar signs Clin Neurol Neurosurg 2011; 113(10): 928-30*
4. *Hamada H, Kurimoto M, Masuoka T, Hirashima Y, Endo S, Harada J. A case of surgically treated acute cerebellitis with hydrocephalus Childs Nerv Syst 2001; 17(8): 500-2*

5. *Horowitz MB, Pang D, Hirsch W. Acute cerebellitis: case report and review Pediatr Neurosurg 1991-1992; 17: 142–5*
6. *Kamate M, Chetal V, Hattiholi V. Fulminant cerebellitis: a fatal clinically isolated syndrome Pediatr Neurol 2009; 41: 220–222*
7. *Perez ER, Maeder P, Cotting J, Eskenazy-Cottier AC, Deonna T. Acute fatal parainfectious cerebellar swelling in two children. A rare or an overlooked situation? Neuropediatrics 1993; 24: 346–35*
8. *Sawaishi Y, Takada G. Acute cerebellitis Cerebellum 2002; 1: 223–8*
9. *Schmucker RD, Ehret A, Marshall GS. Cerebellitis and Acute Obstructive Hydrocephalus Associated with Mycoplasma pneumoniae Infection Pediatr Infect Dis J. 2014;33(5):529-32*
10. *Van Lierde A, Righini A, Tremolati E. Acute cerebellitis with tonsillar herniation and hydrocephalus in Epstein-Barr virus infection Eur J Pediatr 2004; 163(11): 689–91*