



Article review

Acute Cerebellar Ataxia As A Parotitis Complication: Clinical Evaluation And Management

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ABSTRACT

Acute cerebellar ataxia is a motor control or coordination disorder that can be caused by several things, most often in children due to viral infections including paramyxovirus, head trauma, stroke and intoxication with varying prognosis. The paramyxovirus causes parotitis is highly neurotrophic with aseptic meningitis, encephalitis, hydrocephalus, decreased blood flow on the cerebellum and vasculitis. Those mechanisms implicated on many clinical symptoms, including cerebellar ataxia. Management is only limited to supportive and rehabilitative because symptoms will improve within 2-3 weeks.

Keywords : acute cerebellar ataxia, parotitis, clinical evaluation, management

INTRODUCTION

The ataxia is a condition with the main manifestations of loss of the motor control and coordination of the intended movement (Pavone et al., 2017). The other symptoms are the broad spectrum such as speech disorder, abnormal eye movement and difficulty of swallowing (Overby et al., 2019). These varieties of the symptoms depending on the underlying disease and the part of the central nervous system

involved (Ashizawa, Xia, 2016). The incidence of ataxia is still not known indeed, but the incidence of cerebellar ataxia ranges from 30-50% of all ataxia's cases in children with the most common age group is < 6 years old. The proportion of cerebellar ataxia in the pediatric population ranges from 1:100.000-500.000 children (Gilbert et al., 2019; Javadzadeh et al., 2017). The other research conducted in Europe found the results of incidence 26/100.000 children and these results appear to be lower than the global incidence (Musselman et al., 2014). For the cerebellar ataxia which is predominantly hereditary, the incidence 2,7/100.000 and recessive is around 3,3/100.000 children (Pilotto et al., 2018).

Children who have cerebellar ataxia are generally hospitalized because the main clinical symptoms of impaired balance and coordination make these children difficult or unable to do daily activities. Besides that reason, the cerebellar ataxia has many differential diagnoses that must be ruled out to establish appropriate management (Doan et al., 2016). The causes of cerebellar ataxia varied with leading causes are infections including mumps, head trauma, intoxication and cerebrovascular accident (Pedroso et al., 2019; Musselman et al., 2014). The prognosis highly depends on the aetiology of each case. In this regard to this, we would like to make a review of the association of paramyxovirus infections that cause mumps or parotitis with the cerebellar ataxia.

Parotitis or mumps is known as the infectious disease that affects the children with the most common age group 5-9 years old, but this virus can also affect adults. This disease is caused by a paramyxovirus, which is found throughout the world. This disease is quite often found in medical practice so far. From the report states that the national incidence of parotitis 2,2 per 100.000 population annually. Reports from Korea from 2001 until 2015 revealed the increase in the incidence from 10 cases to more than 100 cases/100.000 population annually (Choe et al., 2017). Some case reports suggest that the symptoms of ataxic cerebellar have followed mumps infections in these children (Pedroso et al., 2019). From the literature, we found 1 case report of the neurologic complications after mumps infection (Kang et al., 2014; Uzun, 2015).

Although mumps is known as the self-limiting disease without the specific treatment, some people, this disease made some complications such as encephalitis with all the neurologic sequelae.

PAROTITIS (MUMPS)

Before the era of immunization, mumps was begun, 95% of adult populations were exposed to paramyxovirus (Anderson *et al.*, 1987). After the mumps vaccine carried out in the USA in 1960, the incidence of this disease was sharply declined, and after 1980, the report about mumps disease very rarely. Unlike the conditions in Indonesia, until this day, the mumps is still found in the daily practice. The national incidence of mumps not yet clear, in Ciptomangunkusumo hospital at Jakarta, 61 cases of mumps were found from 1994-1998. However, the records from the other health centre are still few obtained.

From one study conducted in the elementary schools in Jakarta found that the level of knowledge of parents and the socialization of Mumps-Measles-Rubella (MMR) vaccine very minimal. It was proved by from all of the population were include in this study, only 15,7% has the MMR immunization (Satari, Kuniati *et al.*, 2004). The mumps reports and its complications do not report nationally.

CLINICAL EVALUATION OF ACUTE CEREBELLAR ATAXIA

Anamnesis

The anamnesis is intended to search the onset that can be utilized to estimate the aetiology of ataxia. Besides that, the questions that were compiled expected to determine the aetiology of ataxia from peripheral or central problems (Sivaswamy *et al.*, 2014).

- Headache history at frontal/temporal or occipital: basilar migraine or space-occupying lesion in brain stem or cerebellum.
- Headache in children with unknown causes or after falling: swallowing toxic substances or concussion

- Excessive vomiting: basilar migraine, stroke, space-occupying lesion, concussion and benign paroxysmal vertigo
- History of fever with rash and abdominal pain, contact history of infection: Acute-Demyelinating encephalomyelitis (ADEM), acute cerebellitis, Kawasaki disease, Henoch-Schonlein purpura
- Post immunization: Guillan-Barre syndrome or acute post-infectious cerebellar ataxia
- Previous viral infections: acute post-infectious cerebellar ataxia, acute cerebellitis, miller-fisher syndrome
- Nystagmus: benign paroxysmal vertigo, neuroblastoma, Multiple Sclerosis (MS), drug ingestion, stroke
- Head trauma and cervical at adolescent: stroke or concussion
- Seizures: postictal ataxia, drug overdose, fluctuating drug blood levels
- Encephalopathy (impaired consciousness, confusion, aggression, psychosis): ADEM, MS, drug ingestion
- dizziness: benign paroxysmal vertigo, basilar migraine or stroke
- previous episodes of ataxia that self-limiting from family history: episodic genetic ataxia
- history of emotional trauma: psychogenic

Based on the nature of clinical manifestations, ataxia can be arranged the possibility of the underlying aetiology. . Based on the timing of clinical manifestations, we can predict the cause of ataxia beside from the others data like physical examinations. Episodic ataxia (progressively symptoms within minutes to several hours): autoimmune process, genetic, intoxication and vascular problems; acute ataxia (severe symptoms in a few minutes to several days): infections, metabolic disorder, intoxication, trauma capitis and vascular abnormalities; subacute ataxia (symptoms worsen in few weeks to months): autoimmune disease, infections, inflammatory processes, neoplasms and paraneoplastic; Chronic ataxic (the disease develops in several years to decades): autoimmune disease, degenerative disease,

genetic, inflammatory processes, metabolic disease, neoplasm and paraneoplastic; and static ataxia (progression develops in several decades): abnormality of brain development and cerebellar injury (Fogel., 2012).

The incubation period for epidemic parotitis ranges from 14-24 days. The prodromal period is characterized by a feeling of lethargy, pain in the muscles especially the neck muscles, headache, decreased appetite, and is followed by rapid enlargement of one or both of the parotid glands and other salivary glands.

PHYSICAL EXAMINATION

The initial examination should be a focus on identifying conditions that are life-threatening and require immediate intervention (Ashizawa *et al.*, 2016). Neurological examination must be done thoroughly, including evaluation of consciousness both quality and quantity, hallucination, increased of intracranial pressure (pupils, eye movement), nystagmus that may caused by abnormality of vestibular system (peripheral type), evaluations of motoric function to identify weakness of extremities, cerebellum function to identify impairment balance and coordination and sensory and tendon reflexes (Marsden, 2018).

The other physical examinations should be focus to find of the underlying disease as the aetiology of ataxia. In patotitis, we should examined the parotid glands. The enlargement of the gland is accompanied by a feeling of pain and will swell characteristically, starting with filling the space between the back boundary of the mandible and the mastoid bone, then extending in a crescent shape downward and forward, because the upward expansion is limited by the zygomatic bone. The swelling will subside slowly over 3-7 days, but sometimes it can last longer.

PATHOGENESIS AND PATHOPHYSIOLOGY OF ACUTE CEREBELLAR ATAXIA AS PAROTITIS COMPLICATION

Cerebellar ataxia is the pathological conditions due to the cerebellar impairment and associated nerve connections. In connecction with the cerebellum function as the regulator of balance and motor coordinator function (Tanburoglu,

Karatas., 2017). Cerebellum is divided based on phylogenetic and functional criteria, such as vestibulocerebellum regulating eye movement and body balancing by modulating information in the vestibular and reticular nuclei; spinocerebellum which receives peripheral sensory input and regulating body and leg movement and contributes to locomotor, balance and muscle tones; cerebrocerebellum that connected with cerebral cortex and influence movement planning and evaluating sensory information for movement, fine motor, coordinated distal movement, emotion, and cognition. Information of the cerebellum function is essential to know for search the ataxia aetiology and which parts of the cerebellum are affected based on clinical symptoms.

The aetiology of ataxia is quite extensive, based on the onset of symptoms, ataxia is divided into three categories: hereditary, acquired and idiopathic (Tirada, Levy, 2014). The classification of ataxia can also be based on the onset of the disease, acute, subacute and chronic. From those classifications always related to the possibility of the underlying disease.

Acute cerebellar ataxia is mostly secondary after viral infections (enterovirus, Epstein Barr, hepatitis A, herpes simplex, influenza, measles, mumps, parvovirus B19, varicella) and a small portion can be after bacterial infection (*Listeria monocytogenes*, *Legionella* or *mycoplasma*). Both of viral and bacterial infection could be induced encephalitis and cerebellitis (Javalkaret al., 2014) which causes the cerebellar dysfunction which mediated by the autoimmune process (Mitoma et al., 2016).

The role of mumps virus in causing cerebellar ataxia is related to the molecular biology of this virus. Paramyxovirus is the neurotropic virus with the evidence of Central Nervous system (CNS) involvement >50% based on cerebrospinal fluid analytic that reveal the pleocytosis (Rubin et al., 2015; Atluri et al., 2015). The incidence of symptomatic CNS infections is quite rare but significant. Aseptic meningitis accounts for 5-10% of cases and the encephalitis <0,5%. However, in the population have not received MMR vaccination, the percentage of meningitis and encephalitis are increasing. From the previous study, paramyxovirus was identified as

the second most common cause of encephalitis and meningitis (Cantu, M Das, 2019). The description of post mortem pathology consists of brain oedema with bleeding, perivascular lymphocytic infiltration, perivascular gliosis and demyelination. Acute demyelination is caused by a cross-reaction between antibody and epitope in the cerebellum (Burrell, Murphy, 20172).

From the animal models found that the virus transmits into the cerebrospinal fluid through the choroid plexus or mononuclear cell during viremia period. In this site, the virus replicates inside the ependymal cells that lining the ventricle. From this site, the virus enters the cerebral cortex and hippocampus. The ependymal cells that inflammation subsequently degenerated and apart from ventricle wall and entering to the cerebrospinal fluid that may cause the obstruction or stenosis of aqueducts Sylvie and cause hydrocephalus (Wright *et al.*, 2019).

For the objective evaluation, Magnetic Resonance Imaging (MRI) or Computed Tomography (CT) scan is needed to identify the abnormalities caused by a direct or indirect invasion of the virus. From several studies that use the MRI or CT Scanning, reveal demyelination and the other changes due to the inflammatory process (Samkar *et al.*, 2017). A follow-up study was conducted to evaluate cerebral blood flow in patients with cerebellar ataxia after viral infection revealed that mean cerebral blood flow decreased significantly in the cerebellum (Garcia-Iniguez *et al.*). This result indicates the presence of vasculitis or vasculopathy at cerebellar blood vessel while the structure and intensity of cerebellar signal still in normal limits.

Based on the description above it can be said that mumps or parotitis can cause complications in some individuals in the form of demyelinasi and inflammatory processes in the cerebellum. Besides this, mumps also causes vasculitis or vasculopathy of the cerebellar arteries causing ataxia.

MANAGEMENT OF ACUTE CEREBELLAR ATAXIA AFTER PARAMYXOVIRUS INFECTIONS

Management of acute cerebellar ataxia depended from the aetiology and consequences of cerebellar ataxia. In mumps cases, management is just supportive

and rehabilitative. Some health centre does not provide specific therapy because the symptoms will improve within 2-3 weeks after onset. The critical thing is assistance to the child so that physical injuries do not occur due to movement and psychological disturbance. Psychologic mentoring for older children significantly prevent depression caused by the limitation of their activities, although only temporary. The other hand, the consequences of ataxia, must be highlighted in management in these patients. From the literature shown that the children who have the abnormal finding in CT or MRI correlate with neurological disability (Lancella L *et al.*, 2017) and need the specific management from the medical until surgical therapy.

CONCLUSION

Acute cerebellar ataxia causes significant anxiety both in children and their family because of the clinical manifestation cerebellar Ataxia cause limitation of daily activity. The aetiology of this disease vary greatly and must be established by radiology and laboratories examination. Ataxia caused by paramyxovirus does not need specific management.

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