Does Beta Thalassemia Increase the Incidence of Bell's Palsy?

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Abstract: A five-year old Iraqi patient with beta thalassemia major was presented with recurrent attack of Bell's palsy on 2 successive years being a milder one in the second year. This is a rare occurrence in beta thalassemia major patients and needs to be further explored. This is a case report of a nine-year old Iraqi child who is the only child born to parents of secondary consanguinity relatives from Babil Province who was diagnosed as beta thalassemia on the age of 6 months and is on regular blood transfusion and iron chelation therapy of deferasirox tablets (Exjade), his medical follow up takes place at Babil Thalassemia center in Babil Maternity and Child Hospital. On the age of five years, he was presented with a sudden onset of right sided Bell's palsy within twenty four hours of swimming in a cool swimming pool at his home in July 2011. A neurosurgeon examined him at that time, no investigations were done, the diagnosis was purely clinical, steroid & neurotonics were recommended as a treatment for the condition, antiviral agents were not prescribed, lacrimation & taste were not affected, there was a slow improvement, two weeks later physiotherapy sessions including facial massage and electric stimulation were recommended after which the child improved. The next year, another attack of Bell's Palsy developed at a similar time with the same sequel of events, this time the attack was milder and with residual effects seen in the eye & mouth. This coincidence of beta thalassemia and Bell's palsy should be further studied by observational epidemiologic studies to prove or disprove the hypothesis that thalassemic patients especially children are at higher risk of developing Bell's palsy than healthy children. Beta thalassemia patients may be at higher risk of developing Bell's Palsy.

Key words: B Thalassemia major, Bell's palsy, Babil Thalassemia center.

1. Introduction

A five-year old Iraqi patient with beta thalassemia major was presented with recurrent attack of BP (Bell's palsy) on 2 successive years being a milder one in the second year.

This neurological complication in beta thalassemia major patients is rare, this is important to consider in these patients & needs to be further explored to find out whether thalassemic patients are at higher risk for developing BP in order to initiate preventive measures [1].

2. Case Report

This is a case report of a nine-year old Iraqi child who is the only child born to parents of secondary consanguinity relatives from Babil Province, was

diagnosed as beta thalassemia major at the age of 6 months and is on regular blood transfusion and iron chelation therapy of deferasirox tablets (Exjade), his medical follow up takes place at Babil Thalassemia center in Babil Maternity and Child Hospital.

At the age of five years, he was presented with a sudden onset of right sided BP within twenty four hours of swimming in a cold swimming pool at home in July 2011.

The diagnosis was made by a neurosurgeon who happened to be a family friend, no investigations were done, the diagnosis was purely clinical, steroid & neurotonics were recommended, antiviral agents were not prescribed, lacrimation & taste were spared, there was a slow improvement, two weeks later physiotherapy sessions including facial massage and electric stimulation were recommended after which the child improved.

A year later, recurrence of BP developed at a

similar time with the same sequel of events, this time the attack was milder and with residual effects seen in the eye & mouth.

3. Discussion

The relevance of this condition arises from the fact that beta thalassemia major is a chronic haemoglobinopathy that already has its own serious manifestations & complications, as for the case report of this patient who developed BP, it may exert an extra psychological pressure and concern on the patient and his family especially with the residual effects on an obvious part of the body which is the face, a focal point for communication and expression [2].

Bell's palsy in childhood accounts for 90% of facial paralysis. Up to 10% of patients with BP will experience recurrence after a mean latency of 10 years. Knowledge on the course of disease and clinical features of BP in children largely depends on the studies of BP among adults which may not always be conclusive as the course of the disease and outcome may be altered with different types of treatment in children as compared to adults [2, 3].

The incidence of BP in the world varies, in most of the series published incidence oscillates between 11 and 40 cases per 100,000 inhabitants per year although figures as low as 8 and as high as 240 cases per 100,000 inhabitants per year have also been reported. The climate, racial factors, the prevalence of predisposing diseases and the demographic features of the regions studied could be involved in these variations in the appearance of BP (Servicio de ORL, 2005).

The probable cause for the facial nerve palsy in thalassemia is pinching of the facial nerve at one of the narrow points along its course, such as within the facial canal, due to widening of diploic bone [1].

A case of multiple nerve palsies in B thalassemia major was reported in Sri Lanka who developed a lower motor neuron facial nerve palsy on the left side,

together with phrenic nerve palsy on the same side, during the course of the illness, no studies were found that established an association between BP & thalassemia [1].

Exposure to cold prior to the onset of BP which was found in this case, is agreed upon in other studies as a trigger for the development of BP the etiology of which is not well established, however it is not clear that cold has a significant independent effect on risk of disease. Although seasonal variations in BP rates were observed, they were usually lower in summer unlike our case which took place in July [3].

The presentation of the case was typical of BP which was spontaneous acute right-sided (unilateral) facial paresis that took place within 24 hours of exposure to cold and the diagnosis was made by exclusion based on the history, clinical picture & neurological examination, no investigations were done. Bell's palsy is largely a diagnosis of exclusion, but certain features in the history and physical examination help distinguish it from facial paralysis due to other conditions [2, 4].

Evaluation of any therapy for BP is difficult because as many as two thirds of patients have spontaneous and complete recovery thus treatment of BP is still controversial. Many patients begin to improve as early as 10 days after the onset, even without treatment. However, patients who have persistent weakness of varying degrees (16%), involvement of other cranial nerves, or a second episode of palsy require further investigations although reports of children with recurrent BP are uncommon with an incidence of 6.5%, one study revealed. Both persistent weakness & recurrence have occurred in this case [2-4].

Clinicians should provide patients and their families with clear and comprehensible information on the treatment options to facilitate patient understanding and shared decision making, which in turn leads to better patient adherence and outcome [2-6].

Corticosteroid treatment was initiated as soon as the

diagnosis was made, this would improve the prognosis of BP especially if prednisolone is implemented within 24 hours of paralysis and is a strong recommendation that improves likelihood of complete recovery [7-9].

Corticosteroid use is based on the theory that BP is due to some inflammatory process & oedema of the facial nerve. Despite the absence of quality trials supporting steroid use in children, oral steroids may be considered in pediatric patients, the benefits and risks of their use in children with BP need to be better elucidated [4-10].

Antiviral treatment was not used as there was no prove of a viral infection in this case; they are not recommended alone as well a weak recommendation is against the addition of antivirals to corticosteroids for patients with mild to moderate severity. Their admission in the treatment is related to the detection of herpes simplex virus in endoneurial fluid in patients with BP and has implicated the virus in the pathogenesis of the disease. In children, the dose of prednisone and antiviral agents must be adjusted for weight [2, 6, 7].

The patient's family were also advised to do physiotherapy when recovery was incomplete after 2 weeks. There is no recommendation regarding the use of exercise physiotherapy for acute BP of any severity and is suggested for patients with persistent weakness. The prognosis of BP in children seems to be good with higher recovery rate and physical therapy program might be an effective method to facilitate recovery in patients with poor prognosis "Duygu Cubukcu, 2013" [3, 4, 7].

The use of electrostimulation is a weak recommendation as the safety profile of such therapy is unproven as well as there is insufficient evidence to support its effectiveness when applied as monotherapy nor when combined with other procedures [4, 5, 11].

Routine use of surgical decompression is a weak recommendation and patients should consider this option only if they have severe facial nerve

degeneration on electroneuronography, if they are willing to accept the surgical risks and if the surgery is to be performed in an advanced treatment facility.

For a patient with permanent facial paralysis despite medical and surgical treatment, many surgical options are available to improve facial function and appearance [4-6].

4. Results

This coincidence of beta thalassemia and BP should be further studied by observational epidemiologic studies to prove or disprove the hypothesis that thalassemic patients especially children are at higher risk of developing Bell's palsy than healthy children.

5. Conclusions

Beta thalassemia patients may be at higher risk of developing Bell's palsy.

6. Recommendations

Further studies can be done to prove or disprove this hypothesis.

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