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CLINICAL REPORT

A Case Report of Infantile Ramsay Hunt Syndrome

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Abstract Ramsay Hunt Syndrome is a rare condition in children. There are currently no internationally accepted protocols in the management of these patients. We present a case of a 9 month old child that presented to our Department with Ramsay Hunt syndrome. Included is the management of the clinical condition and a brief literature review. Early identification, a high index of suspicion and prompt treatment is required to achieve a good clinical outcome.

Keywords Ramsay hunt · Infant · Cellulitis

Background

Lower motor neuron facial nerve palsy in children may be congenital or acquired. The majority of these may be classified as Bell's palsy however there have been cases reported of pediatric Ramsay Hunt Syndrome (RHS) [1].

Ramsay Hunt was the first to propose that the Varicella Zoster Viral infection of the geniculate ganglion was responsible for the clinical syndrome consisting of facial nerve paralysis, hearing and balance dysfunction, with a herpetic skin rash involving the auricle and thus coined the term ''herpes zoster oticus''[2–4].

The reported incidence is 2.7/100,000 children [5].

The first documented case was reported in a 3 month old by Balatsouras et al. [6].

We present a case of a 9 month old child that developed Ramsay Hunt Syndrome and associated facial cellulitis. We include our management as well as a brief review of the relevant literature.

A search of all articles reporting Ramsay Hunt Syndrome in infants, was performed using the PubMed, Medline, and Cochrane Review Databases, from inception until March, 30 2020.

The detailed keywords used for the search were as follows: "Infant", "Ramsay Hunt Syndrome", "Herpes Zoster Oticus".

Case Presentation

A 9 month old child was referred to the Otorhinolaryngology department at the Charlotte Maxeke Johannesburg Academic Hospital by the pediatric department, with a one week history of having chicken pox.

The child was not vaccinated against chicken pox. There was no history of Zoster infection reported by the mother during her pregnancy, and for the following 9 months after the baby was born.

Upon further clinical enquiry it was found that the child had vesicles on the right ear that had burst open.

The mother also noticed a discharge from the right ear and associated swelling over the ear that was progressively getting worse. There was also a swelling on the right side of the face.

The right external ear canal was swollen and on initial examination it was difficult to visualise the tympanic membrane. Vesicles were noted on the Right pinna (Fig. 1).

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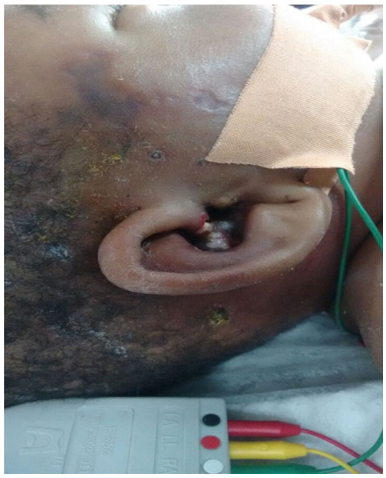

Fig. 1 Herpetic vesicles on the ear and face

The child also had a complete Right peripheral facial nerve palsy (House Brackman 6) (Fig. 2).

A clinical diagnosis of Ramsay Hunt Syndrome was made and the child was admitted for intravenous therapy.

She was started on intravenous acyclovir (10 mg/kg/day), methylprednisolone (1 mg/kg/day) and ceftriaxone (100 mg/kg/day) for a week.

Haematological investigations revealed a white cell count of 25, a haemoglobin of 11.7 and a C-reactive protein of 85. Blood cultures were also done but they proved to be negative after a week.

A CT scan revealed a Right sided facial, scalp, pinna and pre-septal soft tissue swelling.

No soft tissue fluid collections were seen. The right mastoid air cells and middle ear cavity were opacified with no obvious bone erosion. The inner ear structures appeared normal bilaterally.

No focal intra-parenchymal lesions were noted. Normal cerebral parenchyma was found.

Otoacoustic emission (OAE) tests were conducted as part of the audiological assessment, the right ear was deemed as pass, and the left referred due to the middle ear pathology. A decision was taken to treat the middle ear pathology and repeat the hearing tests.

A lumber puncture was done as part of the septic workup. This came back as normal.

The facial swelling subsided within a week and the facial nerve palsy resolved as well (Fig. 3).

Follow up OAE and tympanometry were normal. The child was thus discharged from hospital, with regular follow-up at the ENT clinic. A year later the child was reviewed with no evidence of complications.

Fig. 2 Right facial nerve palsy

Literature Review

Ramsay Hunt syndrome is rare in children and has mainly been documented as case reports.One of the largest case series originates from Japan by Hato et al. who looked at a large-scale study of 311 children with facial palsy. The authors reported that 52 of these patients were affected by Ramsay Hunt syndrome [7].

Varicella Zoster reactivation has been implicated as the pathogenetic mechanism for Ramsay Hunt syndrome in both children and adults, however infection in infants is probably due to a different mechanism that is not fully understood at this stage [8].

Infantile herpes zoster has been reported in the literature, often involving the spinal nerves. In a study involving 51 cases the location was as follows: 14 (27%) were in cranial nerve, 10 (20%) in cervical, 17 (33%) in thoracic, and 10 (20%) in lumbosacral dermatomes. There were cases where more than one dermatome was affected [9–12].

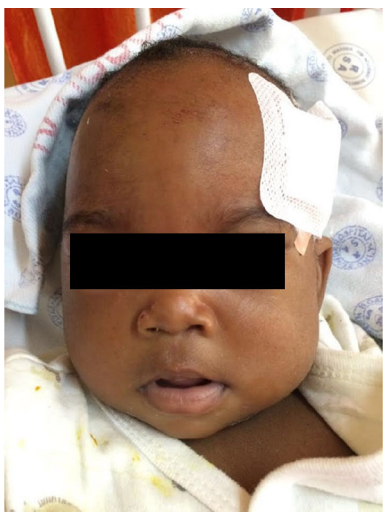

Fig. 3 Child post medical treatment

RHS commences with the classical prodromal phase presenting with deep ear pain, fever, and fatigue, that usually lasts 1–3 days in duration.

Thereafter herpetic vesicles begin to develop in the external auditory canal, tympanic membrane, and/or the anterior two-thirds of the tongue. The facial nerve paralysis develops within 1–2 weeks after the appearance of the herpetic rash [13, 14].

Serological testing by measuring serum anti-VZV IgG and IgM antibody titers using ELISAs may be done, however RHS remains a clinical diagnosis.

This condition is potentially hazardous as complications may include meningitis, brainstem encephalitis and death, hence an accurate diagnosis and prompt treatment is needed. Early commencement of antiviral and high-dose steroid therapy has become the standard treatment for patients diagnosed with RHS at many institutions.

Antiviral agents inhibit viral replication,afford rapid lesion healing and restrict new lesion formation [15].

Double blind controlled studies of oral acyclovir therapy for varicella in healthy children showed no effect on improving immunity, however there have been case reports that have used oral acyclovir. Antiviral agents may inhibit the replication and spread of the virus in the facial nerve and thus prevent further damage if initiated early [8, 16]

We thus felt that this case did merit the use of intravenous antiviral therapy as benefit of therapy far outweighed risk in this case. Commencement of an antiviral agent within the first 72 h is important if treatment is to be effective [8, 17–19].

In patients with RHS, steroids reduce oedema and pain by countering inflammation in the peripheral neurons.

Combined antiviral and steroid treatment has been suggested to be more effective than steroids alone.

Conclusion

Ramsay hunt Syndrome is a rare presentation in children and requires identification and treatment. The mechanism of infantile Ramsay Hunt syndrome may be different to older children and adults. There are still many facets of the disease including the transmission, pathogenesis and treatment modalities that need to be explored through ongoing research.

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Compliance with ethical standards

Conflict of interest There is no conflict of interest.

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