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Case Report

A Congenital Insensitivity to Pain with Anhidrosis (CIPA) in 1.5 Year Old Male

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Abstract

Congenital insensitivity to pain and anhidrosis (CIPA) it is a rare genetic syndrome also known as hereditary sensory and autonomic neuropathy type IV. Patients usually presented with: insensitivity to pain, inability to sweat (anhidrosis), various degrees of mental retardation, osteomylitis, deformities of joints that may lead to fractures. Also these Patients have the ability to severely injure themselves repeatedly which may result in permanent damage and often their condition is misdiagnosed as child abuse. We report a case of 1.5 years old male presenting with this type of syndrome.

Keywords

CIPA, Self Mutilation, Pain Insensitivity.

Declaration of Conflicting Interest

The authors declare that they have no conflict of interest.

Introduction:

Congenital insensitivity to pain and anhidrosis (CIPA) is an autosomal recessive rare genetic disorder caused by mutation in the NTRK1 gene, which encodes tyrosin receptor (TrkA) for nerve growth factor (NGF) (2,3). It is also known as hereditary sensory and autonomic neuropathy type IV. The insensitivity and the presence of anhidrosis in these patients are caused by the absence of NGF-dependant primary afferents and sympathetic postganglionic neurons, respectively (3). Worldwide few hundreds of cases of CIPA have been published (4). The incidence of this syndrome occurs in 1 in 125 million newborns (3).

Some clinical, radiographic and laboratory examinations for these patients may reveal normal blood, urine and cerebrospinal fluid (3). Dislocation of the joints and deformities as well as osteomylitis. in addition to that some oral ulcerations and infections, corneal scarring and opacities can also be found (1,5).Some patients may also show immunological abnormalities such as: abnormal neutrophil function, defect in lymphocyte signaling and humoral immunodeficiency. Additionally we can find some patients suffers from recurrent ***staphylococcus aureus*** infections of bones and skin due to impaired neutrophil chemotactic activity (3,4,5).

Case report

1.5 years old male patient of a Sudanese nationality was the first child of consanguineous marriage (first degree cousins), he wasn't diagnosed with congenital insensitivity to pain and anhidrosis (CIPA) till the time he visited our dental center. He came complaining of sever lower lip ulceration and mobility of the maxillary central incisors as a result of constant subconscious chewing of the lower lip. According to his mother his primary teeth started to erupt at age of 4month and he used to suffer from recurrent fever, infections and tongue ulceration and he was seen by an oral maxillofacial surgeon (OMFS) and he extracted his lower teeth due to the severe mobility.

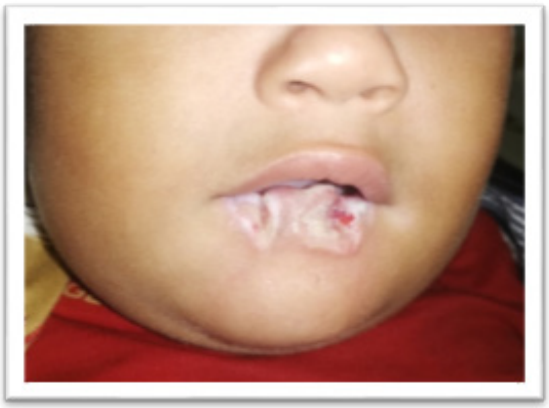


Figure (1): severe lower lip ulceration

Oral gel containing hyaluronic acid was prescribed to aid in the healing of the ulcer.

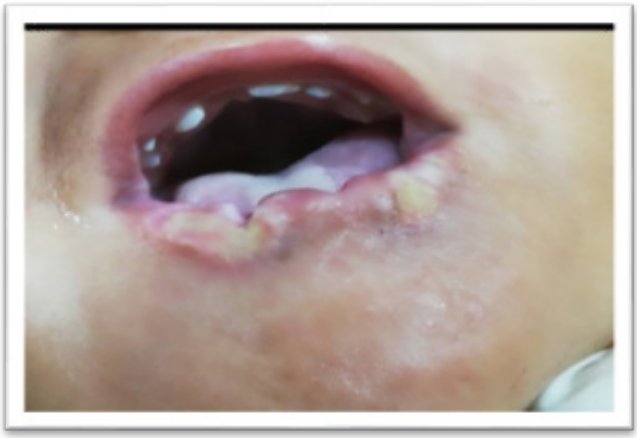


Figure (2): lower lips after few weeks of the usage of oral gel

After few weeks from the 1st visit extraction of the maxillary central incisors was done as well as grinding of the tips of his canines to prevent further damage because the patient did not stop mutilating his lips. Follow up is done and the result was up to the parent's satisfaction.

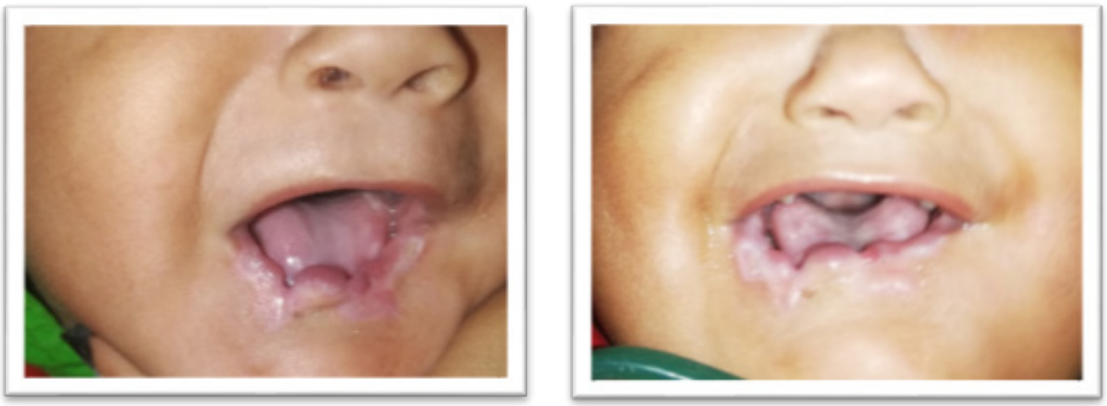


Figure (3): lower lips after the extraction of the upper teeth

Parents were told that he might need lip reconstruction with a plastic surgery later in life. Injury to the patient fingers was also noticed, it shows on figure (3).

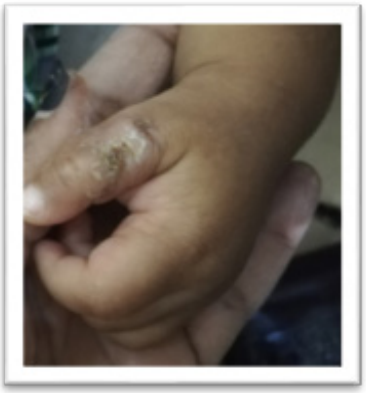


Figure (3): injury to the fingers (thumb)

Tongue scar was seen, plus premature eruption of the primary molars were noted, it shows in figure (4).

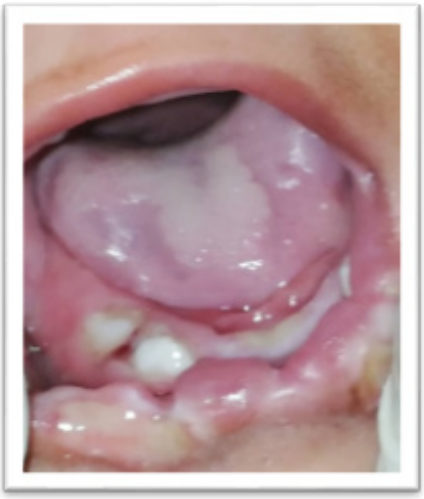


Figure (4)

Few month later the patient came with an extra-oral swelling on the lower right side of the mandible on examination there was a non healing socket of the tooth 83 and 85 with foul odor. His mother said they fell sooner after he bit on a toy very hard he was given antibiotics but the swelling did not decrease in size and it became hard. She brought the fallen teeth with her and they were still immature.

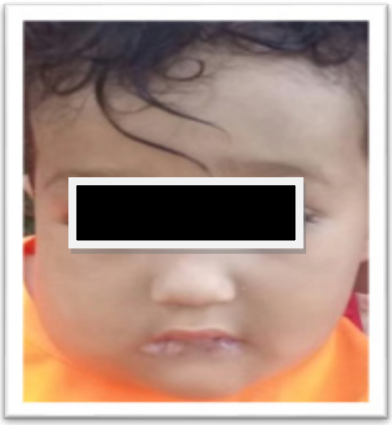


Figure (5): swelling on the lower right side of the jaw

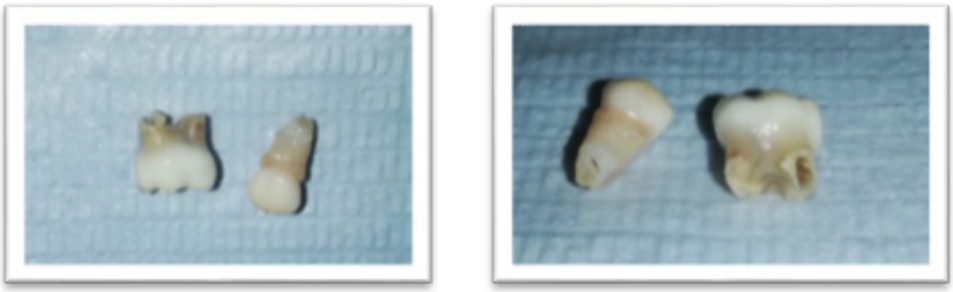
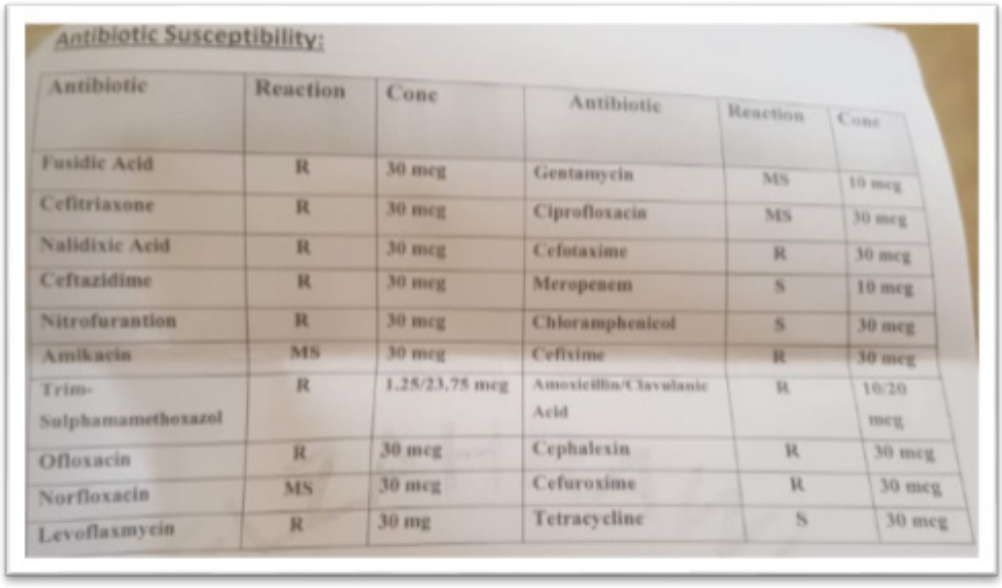


Figure (6): fallen teeth with immature roots.

A swab was made for culture and sensitivity and the microorganisim associated was found to be *klebsiella* species. The patient was resistant for most of the antibiotics due to the misuse by the parents and doctors hence the patient had recurrent fever and infections most of the time.



Antibiotic Susceptibility:

Antibiotic	Reaction	Conc	Antibiotic	Reaction	Conc
Fusidic Acid	R	30 mcg	Gentamycin	MS	10 mcg
Cefitriaxone	R	30 mcg	Ciprofloxacin	MS	30 mcg
Nalidixic Acid	R	30 mcg	Cefotaxime	R	30 mcg
Ceftazidime	R	30 mcg	Meropenem	S	10 mcg
Nitrofurantion	R	30 mcg	Chloramphenicol	S	30 mcg
Amikacin	MS	30 mcg	Cefixime	R	30 mcg
Trim-Sulphamamethoxazol	R	1.25/23.75 mcg	Amoxicillin/Clavulanic Acid	R	10/20 mcg
Ofloxacin	R	30 mcg	Cephalexin	R	30 mcg
Norfloxacin	MS	30 mcg	Cefuroxime	R	30 mcg
Levoflaxmycin	R	30 mg	Tetracycline	S	30 mcg

Figure (7): sensitivity of the bacteria to antibiotics.

Also a CT was made for the facio-maxillary bones and the results found were:
- Destructive changes involving the body and the ramus of the mandible.
- Contra lateral side appears normal
- Normal maxillary bone
- Normal size and attenuation of the TMJ

Simultaneously the patient was complaining of right knee swelling so he was referred to the orthopedic department and knee-leg radiograph was made. Findings suggested presence of chronic osteomylitis of the bones.

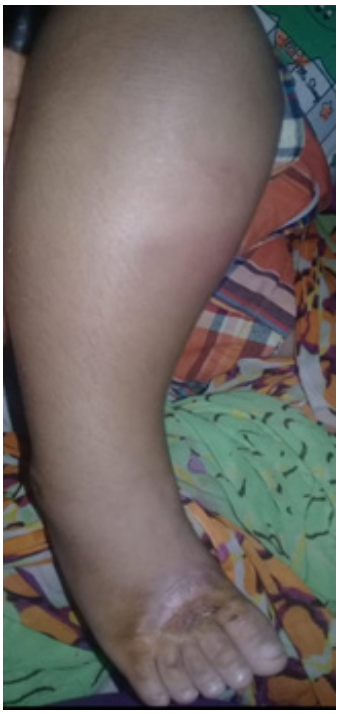


Figure (8): swelling involving the right knee

We can also notice an injury of the foot and some of the toes; it shows on figure (8).

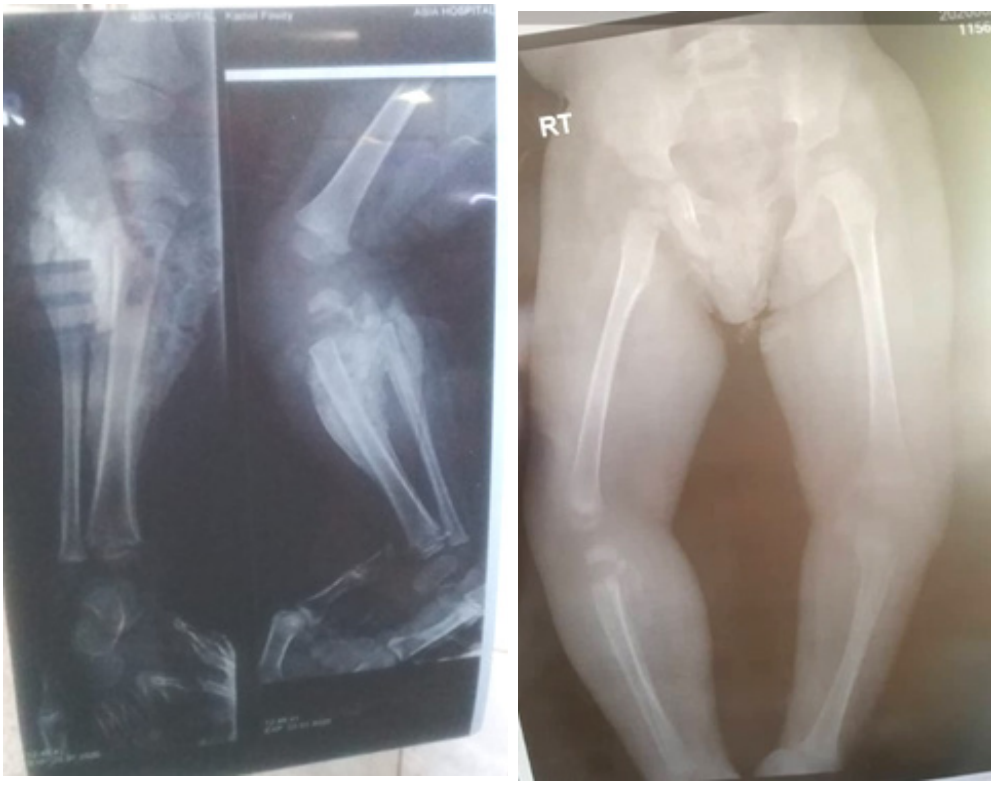


Figure (9): knee-leg radiographs

Discussion:

Congenital insensitivity to pain and anhidrosis (CIPA) can be easily misdiagnosed with child abuse because it has much higher occurrence than rare neuropathies like this one (1).

Diagnosis of such a condition is usually clinically when the patient is presented with impaired pain and temperature perception which will be presented as recurrent episodes of fever which is responsible of death of almost 20% of children (6). Diagnosis of this condition is usually around the age of 3 years but it can be as early as this patient's age, because it might not be well known in the medical community and can easily be missed.

Parents were seeking medical help since he was 4month and they reached the final diagnosis by age of one year and a half after they visited our center. The child was subjected to so many laboratory tests to justify his recurrent fever and infections, though these patients will be presented with these symptoms usually. Examples of these tests are: complete blood picture, immunoglobuline levels (IgA, IgM, IgG), malaria, C-reactive protein level, herpes simplex virus infection, rubella, toxoplasmosis, cytomegalovirus, liver function tests and kidney function test. As well as multiple radiographs.

Marked anhidrosis is seen in those patient and this will disrupts the thermoregulation of the body and can explain the recurrent episodes of fever, this may cause hyperpyrexia and it might be fatal (3).

Regarding treatment of this condition is still not well established but there is some drugs like: naloxon, can be used to reverse the analgesia (4). Other options of therapy can be limited to prophylactic measures and treating patients according to their symptoms (5). Also prevention from infections may improve their condition, specially designed shoes may prevent them from foot injuries and avoid surgeries early in life (6)

Treating such patients requires experience, knowledge and collaborative team of medical professionals.

Finally, providing education, care and support for those patients and their parents is necessary to maximize the quality of their lives (1, 2).

Conclusion:

There is no specific or gold standards treatment of this condition; it is a congenital disease that results from gene mutation. So the early diagnosis and treatment of the infectious conditions may help in managing the case from an early age and prevent life threatening catastrophic complications later in life.

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