ejpmr, 2018,5(8), 490-491

SJIF Impact Factor 4.897

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

CASE REPORT ON INCREASED RISK OF CELLULITIS IN PRADER WILLI SYNDROME (PWS)

Aisha Jalaludeen* and Said Ali Muhammad

Doctor of Pharmacy Interns (Pharm-d) Sree Krishna College of Pharmacy, Trivandrum.

***Corresponding Author: Aisha Jalaludeen**

Doctor of Pharmacy Interns (Pharm-d) Sree Krishna College of Pharmacy, Trivandrum.

Article Received on 28/05/2018 **Article Revised on 18/06/2018** **Article Accepted on 08/07/2018**

ABSTRACT

Prader-Willi syndrome (PWS), first described by Prader, Labhardt and Willi in 1956, is a congenital disorder in which various configurations of seven genes on chromosome 15. Prader-Willi syndrome is a complex disorder affecting multiple systems with many manifestations relating to hypothalamic insufficiency. Major findings include infantile hypotonic, developmental delay and mental retardation, behavior disorder, characteristic facial appearance, obesity, hypogonadism, and short stature. Obesity and the behavioral problems are the major causes of morbidity and mortality. In our case study we discuss about a 27 year old male patient who is a known case of Prader Willi syndrome with morbid obesity since 2009 and now suffers from Extensive Filarial Cellulites right lower limb. He is also a known case of Type 2 respiratory failure.

INTRODUCTION

Prader-Willi syndrome is a rare genetic disorder that results in a number of physical, mental and behavioral problems. A key feature of Prader-Willi syndrome is a constant sense of hunger that usually begins at about 2 years of age. People with Prader-Willi syndrome want to eat constantly because they never feel full (hyperphagia), and they usually have trouble controlling their weight. Many complications of Prader-Willi syndrome are due to obesity. Best managed by a team approach, various specialists can work with you to manage symptoms of this complex disorder, reduce the risk of developing complications and improve the quality of life for your loved one with Prader-Willi syndrome Prader-Willi syndrome is a genetic disorder, a condition caused by an error in one or more genes. Although the exact mechanisms responsible for Prader-Willi syndrome haven't been identified, the problem lies in the genes located in a particular region of chromosome 15.

Prader-Willi syndrome occurs because certain paternal genes that should be expressed are not for one of these reasons:

- Paternal genes on chromosome 15 are missing.
- The child inherited two copies of chromosome 15 from the mother and no chromosome 15 from the father.
- There's some error or defect in paternal genes on chromosome 15

Signs of cellulitis can be difficult to ascertain in the very obese individual since the legs are often already chronically swollen, indurated (skin thickened and hard) and discolored even without infection.

Note

1. Pitting is usually absent even in the presence of massive oedema
2. Abnormalities on chest radiograph (X-ray picture) and even echocardiography are a very late finding long after the individual is quite compromised.
3. End stage obesity hypoventilation includes right heart failure from pulmonary hypertension and, far less often, some left ventricular dysfunction. For this reason, despite massive tissue oedema, pulmonary oedema is not typically part of the clinical picture. Increased activity is more effective and safer than diuretics.
4. Cellulites: Abnormalities in white blood cell count, fever, pain or measurements of inflammatory markers can be delayed or minimal even with serious infections. Oral antibiotics, sometimes in combination with an antifungal agent (such as fluconazole) are usually effective.

In Patients with PWS. Examination of the legs by caretakers seeking changes in feel or appearance is essential.

Patients do not always exhibit fever or pain. Limited cases of cellulites, diagnosed early, can be managed with oral antibiotics. Preventative use of antibiotics is discouraged to prevent development of resistant strains of bacteria. However, intravenous antibiotic may be necessary in severe cases, especially if there is evidence of systemic infection (fever or malaise). In all cases, an attempt should be made to identify the causative

organism but this is not always possible. Maintaining and increasing physical activity and leg elevation when the patient is sitting have proven useful adjuncts in the management of these difficult conditions. Cellulitis and superficial venous thrombosis are not reasons to limit activity; rather, the reverse is true. Patients who have ceased to walk for any reason are at high risk for thromboembolic events (blood clots) and prophylactic anticoagulation should be considered.

CASE REPORT

A 27 year old male patient who is a known case of Prader Willi Syndrome was admitted in a tertiary care centre with complaints of fever since a day, swelling of right lower limb and now complaints of swelling with oozong since a day. On local examination it was found out that there was redness over the entire right lower limb with lymphangitis.

His laboratory findings showed an elevated total count - 27250 cells /µ L. He was admitted and the following was his treatment plan.

Inj.Piperacillin Tazobactam 4.5g IV Q8H
Inj.Moxifloxacin 400 mg IV OD
T.Linezolid 600 mg 1-0-1
T.Alanz (Trypsin+Bromelain+Rutoside) 1-1-1
T.Diethylcarbamazine 100 mg 1-1-1
T.Rabeprazole 20mg 1-0-1
T.Cobadex CZS (Multivitamin)
T.Paracetamol 650 mg 1-0-1
GM wound dressing to lower legs

The above treatment plan was continued for 4 more subsequent days during his hospital stay and on day 4 the superficial blisters deroofed completely and Total count was normal 8570 cells /µL. He was discharged with the following drugs on the 5th day when his redness and edema subsided and his cellulitis resolved.

T.Cefdinir 300 mg 1-0-1 x 3 days
T.Rabeprazole 20 mg 1-0-1 x 3 days
T.Cobadex CZS 1-0-0 X 1 week.
T.Diethylcarbamazine 100 mg 1-1-1 x 1 week

CONCLUSION

Overuse of oxygen and of diuretics are the 2 most dangerous and common errors of rehabilitating persons with obesity hypo-ventilation and edema.

Even persons who are very compromised from their obesity and low oxygen levels must begin to move even if it is only a few more steps per day than previously attempted. Using a walker is very helpful for the sick patient who has been very sedentary.

Using lots of encouragement and incentives, persons with PWS can be induced to walk more each day with a goal of 1 hour per day (in 2 shorter sessions). They will virtually always experience a great improvement in their edema. If the leg swelling has been present for years some swelling of the lower legs will often remain due to the damage to the veins and lymphatics. Therefore early Intervention is ideal.

Most adults with Prader-Willi syndrome are unable to live fully independent lives, such as living in their own home and having a full-time job because their behavioural issues and problems with food mean these environments and situations are too demanding. However, adults with Prader-Willi syndrome can have active social lives and become involved in clubs or volunteering. Adults with the syndrome who don't live with their parents will probably require residential care.

REFERENCE

1. The Journal of Clinical Endocrinology & Metabolism, 1 November 2012; 97(11): 3837–3844.
2. Ledbetter DH, Riccardi VM, Airhart SD, Strobel RJ, Keenen SB, Crawford JD. Deletions of chromosome 15 as a cause of the Prader-Willi syndrome. N Engl J Med., 1981; 304: 325-9.
3. Cassidy SB. Prader-Willi syndrome. Curr Probl Pediatr, 1984; 14: 1-55.
4. Lee PDK. Endocrine and metabolic aspects of Prader-Willi syndrome. In: Management of Prader-Willi syndrome. New York: Springer-Verlag, 1995; 32-57.
5. Dykens EM, Cassidy SB. Prader-Willi syndrome: genetic, behavioral and treatment issues. Child Adolesc Clin N Am., 1996; 5: 913-27.
6. S. T. Lo, P. J. L. Collin and A. C. S. Hokken-Koelega, Visual-motor integration in children with Prader–Willi syndrome, *Journal of Intellectual Disability Research*, 2015; 59(9): 827-834.
7. J. Sloneem, K. Arron, S. S. Hall and C. Oliver, Self-injurious behaviour in Cornelia de Lange syndrome: 2. association with environmental events, *Journal of Intellectual Disability Research*, 2009; 53(7): 590-603.