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Case report - moderate covid 19 diseases in a patient with jarcho levine syndrome

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Abstract

Jarcho Levine Syndrome (JLS) is a rare congenital illness with no distinct gene marker. It is characterized by short trunk dwarfism, a petite neck, chest and rib ab normalities, a kyphoscoliotic curve, and other vertebral anomalies. 2 Although the exact genetic cause of JLS syndrome has not yet been identified, MESP2, LFNG, HES7, and TBX6 have frequently been implicated. Treatment includes aggressive neonatal management, asepsis, and planned orthopedic procedure. We present a case of 2-year female child with Jarcho levine syndrome who presented with moderate covid 19 disease.

The child presented to our tertiary care centre with high grade fever, cough, respiratory distress. On admission, the child was having hypoxia with audible grunting and altered sensorium. Covid 19 RTPCR was positive and the child was classified as having moderate covid 19 diseases. She was put on High flow nasal cannula, Intravenous fluids, 3rd line iv antibiotics and Methyl prednisolone pulse therapy. Chest x-ray was suggestive of bilateral paracardiac pneumonia and crab like rib anomaly. Blood investigations suggestive of raised in flammatory markers Procalcitonin, interleukin 1, and ferritin.

The patient gradually improved over 2-3 days and was successfully discharged at 10th day of admission.

Keywords: Khyposcoliotic, Covid 19, Orthopaedic, Crab Like Rib, Pneumonia, Hypoxia, Antibiotics.

Jarcho Levine syndrome is a congenital skeletal disorder with an autosomal recessive inheritance pattern.

JLS incorporates two separate genetic disorders, spondylocostaldyostosis[1] and spondylothora icdysplasia characterized by segmentation defect of vertebra including hemivertebra, butterfly vertebrae. Malformed small chest of these children predisposes them to reduced lung capacity, recurrent respiratory infections, respiratory difficulty with covid 19 infection further com plicating their clinical course.

Here we present a JLS patient who presented to our tertiary care center with moderate covid 19 illness.

Case report

A 2 year old female child presented to our tertiary care center, with complains of high-grade fever with multiple spikes for last 4 days along with dry, paroxysmal cough with nocturnal exacerbation. Patient was being treated by some OPD based medication earlier given by private hospital however, the child developed progressive respiratory distress in form of severe tachypnea, indrawing of lower chest, reduced oral intake and was transferred to our care.

On admission, the child was having high fever and severe respiratory distress with audible grunting along with altered sensorium (GCS 10/15).

On clinical inspection the child had a short stature, short trunk with scoliosis. Misaligned “crab like” rib cage in form of Pectus Carinatum and protuberant abdomen visualized. The child had lumbosacral unruptured men Ingo myelocele with isolated gross motor delay.

On auscultation, diffuse bilateral crepitations were heard and loud heart sounds were present.

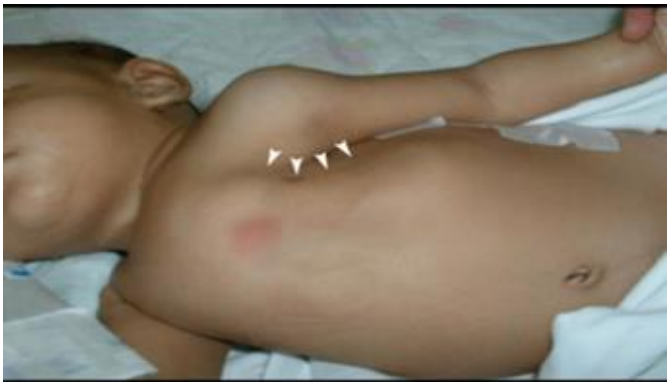

Fig1: Pectus Carinatum

In view of the above clinical picture, during the covid pandemic, covid 19 RTPCR was done, which turned out to be positive putting a major threat on the child, who already was having Restrictive pattern of lung injury, associated with J-L syndrome.

The challenging task of salvaging the child was undertaken. According to MOHFW guideline the child with fever, altered sensorium and severe tachypnea, was classified as having moderate covid 19 disease. She was put on High flow nasal cannula, Intravenous fluids for hydration. Initially, she was having a borderline spo2 of 85-89%.

3rd line iv antibiotics and Methylprednisolone (1mg/ kg/ day) pulse therapy was started as per protocol. Chest x-ray was, suggestive of bilateral paracardiac pneumonia along with crab like rib anomaly hallmark of the syndrome. Blood investigations showed raised inflammatory markers- Procalcitonin, interleukin 1 and ferritin. Electrolyte, 2D Echo and ECG turned out to be normal.

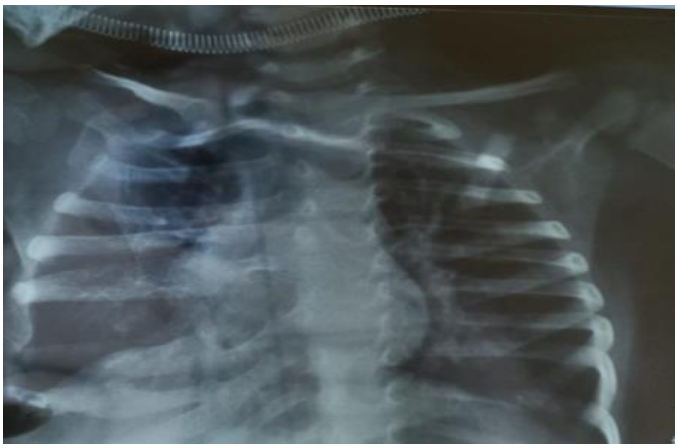

Fig 2: CXR suggestive of crab like ribs

Responding to the treatment of covid 19, the patient gradually improved over 2-3 days, tapered off oxygen support and was successfully discharged at 10th day of admission.

Discussion

The prevalence of Jarcho Levine Syndrome, a rare congenital illness, is unknown because the majority of cases go undetected or misdiagnosed. [2] Although the

exact genetic cause of JLS syndrome has not yet been identified, MESP2, LFNG, HES7, and TBX6 have frequently been implicated. Clinically, the patient is immobile due to short trunk dwarfism and a petite neck. Chest and rib abnormalities increase the likelihood of recurrent pneumonia of varied severity and hinder lungs from developing. According to imaging results, the spine has a kyphoscoliotic curve and is segmented into at least 10 adjacent levels by vertebral anomalies including hemivertebra and butterfly vertebra. The characteristic "Crab Rib"[3,4] appearance is caused by misaligned, packed ribs with fusion at the posterior costovertebral joint and flared anteriorly. There is no distinct gene marker for JLS, hence detailed family personal history and clinical examination helps in stamping the disease. In utero diagnosis can be suspected in view of spine and chest defects. Treatment [5]of the JLS depends on degree of skeletal deformity and thoracic insufficiency. Aggressive neonatal management in form of respiratory support, asepsis along with planned orthopedic procedures at later stage.

Management of Pediatric Covid 19 guideline:[6,7]

1.Assymptomatic: incidentally detected, suspected contact

- Home isolation
- No specific medication required for covid 19 infection.
- Ensure fluids and feeds. Continue medication for any other cause.
- Vital monitoring and use masks.

2. Mild: sore throat, cough without breathing difficulty, spo2 more than 94% on room air

- Symptomatic treatment for fever, cough
- Ensure fluids and feeds, vital monitoring, isolation, hand hygiene.
- Antimicrobials and additional tests not needed.

3. Moderate in addition to fever cough sore throat, rapid breathing as per age of the child, spo2 90-93% on room air

- Oxygen supplementation
- Fluid and electrolyte balance. Antipyretics to be used
- antimicrobials

4. Severe spo2< 90% on room air, any of the following: signs of severe respiratory distress, septic shock, multiorgan dysfunction, seizure, somnolence.

- Initiate oxygenation, fluid and electrolyte balance
- Corticosteroids to be given
- Anticoagulants may be needed
- Antimicrobials

Conclusion

JLS is a potentially chronic debilitating congenital anomaly. Though with modern imaging and orthopedic surgical interventions have improved the survival in these children but those who survive beyond early childhood suffer progressive scoliosis, nerve compression, paraplegia etc. Moreso these children had been exposed to risk of life-threatening respiratory illness in covid era due to inadequate vaccine coverage as also increased covid 19 susceptibility in them.

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