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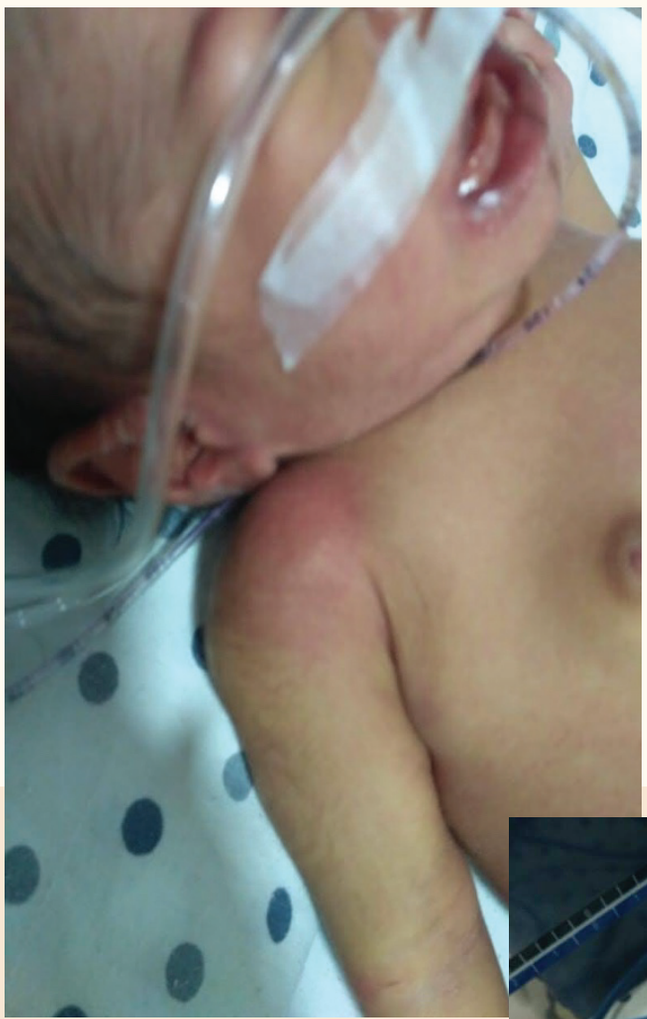
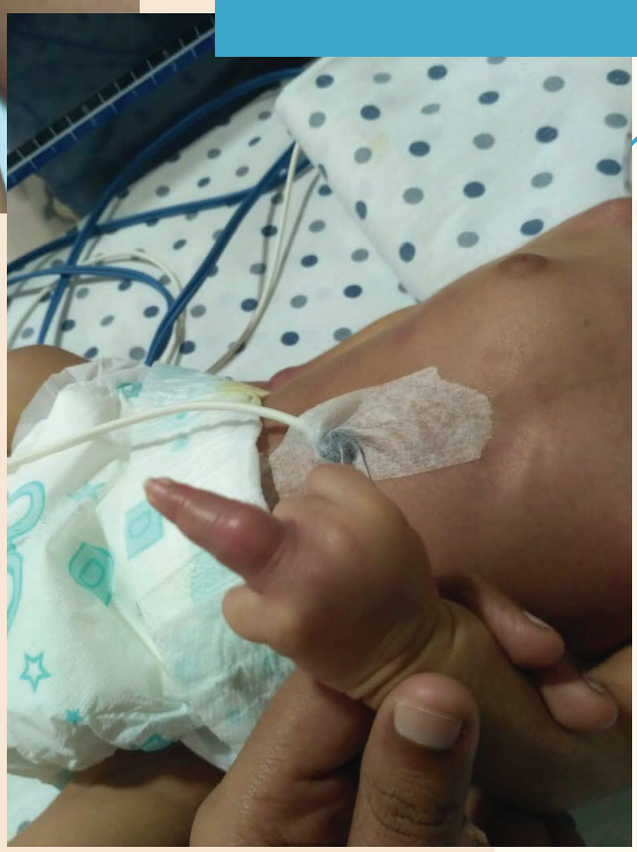


MEDICAL INSIGHTS

EDITION-10



CASES:

www.ankurahospitals.com

Founder’s Note

Greetings from Ankura!

2022 is coming to an end, and as everyone looks towards a new year with promise and gusto, we are excited to unveil an invaluable gift for Hyderabad. Presenting our newest entrant into the Ankura family: 9M by Ankura Hospitals, a specialty ob-gyn facility that will redefine healthcare for the modern woman!

This is a historic moment for our organization as we venture into a more focused approach for the women of our community. 9M by Ankura Hospitals will be a modern institution that boasts of the best healthcare and ob-gyn professionals, the very latest technology and advances in healthcare for women. It also brings the complete healthcare experience for women and children that we are known for – under one single roof!

Dr. Krishna Prasad Vunnam

Founder & MD
Ankura Hospital for Women & Children

I would like to thank our leadership and operations teams for making the new centre a reality for the communities in Gachibowli and surrounding areas. The journey of how far we’ve come along and conceived 9M is special to me, and we’re even more excited to see where this journey takes us into the future. 2022 will see us stepping outside the Telugu states for the first time, heralding a national footprint!

The past few years have been unprecedented for society, and the patience and faith our medical fraternity has displayed is truly inspiring! Healthcare is equated to God’s work, and we shall leave no stone unturned to become the most sought-after healthcare provider for women and children in India. As we embark on this new journey, I look forward to your support. Here’s wishing you a happy and healthy 2022!

Best Wishes,
Dr Krishna Vunnam

Message From The Editor

Editor
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Dear friends,

Immunodeficiencies (primary and secondary) are very important conditions that can present and affect our patients in various ways. To diagnose the conditions early and prevent complications, paediatricians need to watch out for the warning signs of primary immunodeficiency conditions (PIDs). Traditionally the ten warning signs for PIDs were developed for alerting the public and clinicians about the possibility of PIDs. They include:

- Four or more new ear infections within 1 year
- Two or more serious sinus infections within 1 year
- Two or more pneumonia within 1 year
- Two or more deep-seated infections including septicaemia
- Recurrent deep skin or organ abscesses
- Persistent thrush in the mouth or fungal infection on the skin
- Two or months on antibiotics with little effect
- Need for intravenous antibiotics to clear infections
- Failure of an infant to gain weight or grow normally
- A family history of a primary immunodeficiency

These warning signs emphasize predominantly recurrent/persistent infections. However, as our understanding of PIDs is increasing rapidly, PIDs now include diseases that present with sporadic infections, autoimmunity, autoinflammation, and malignancy.

Ankura hospitals are proud to be associated with two renowned pediatric immunologists Dr. Sagar Bhattad and Dr. Anjani Gummadi. With their help, we have been able to diagnose and treat various PIDs and improve the quality of life of our patients. We hope that this edition will increase the awareness of our readers regarding this very important condition.

Message From The Guest editor

Guest editor
Dr. Anjani Gummadi
MD (Pediatrics), PGI, Chandigarh,
DM (Pediatric Clinical Immunology and Rheumatology), (PGI, Chandigarh)
Consultant Pediatric Immunologist & Rheumatologist
Ankura Hospital for Women & Children

Dear friends,

Pediatric rheumatology and immunology is an rapidly evolving clinical discipline which deals with diverse group of systemic disorders. These disorders include juvenile arthritis, connective tissue disorders like systemic lupus erythematosus, dermatomyositis, vasculitic disorders like Kawasaki disease, Henoch schonlein purpura, autoinflammatory disorders and various forms of inborn errors of immunity.

This specality requires a diagnostic approach to make a rapid and appropriate diagnosis which needs a multidisplinary involvement of various subspeciality of Pediatrics. Treatment of rheumatological and immunological disorders in children is challenging and requires long term follow up.

There is poor awareness on this group of disorders leading to delay in diagnosis causing morbidity and mortality. Hence, in this edition we bring to you 4 interesting cases: 2 cases of inborn errors of immunity with different presentations i.e eczema and severe infections and 1 cases of multisystem inflammatory syndrome in neonates and one case of arthritis.

Symptoms which indicate an underlying rheumatological disease in children

- Fever: persistent, recurrent, prolonged, unexplained, Not responding to antibiotics
- Swelling and stiffness of joints
- Joint pain
- Change in gait, limp
- Pain or weakness in the muscles
- Fatigue/extreme weakness
- Skin Rash
- Oral or genital Ulcers
- Dryness of mouth, skin tightening
- Unexplained Hair loss
- Redness of eyes, decreased vision
- Loss of weight, Loss of appetite

Consider underlying immunodeficiency

1. Infections are severe enough to cause hospitalisation
2. Infections which are difficult to treat and do not show response to treatment
3. Infections which are recurrent
4. Infection with unusual organism
5. Infections at unusual site
6. Infections/diseases similarly in family members

Contributors

Topic: An Unusual Cause Of Severe Eczema

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Topic: Infantile Onset Arthritis: Not Always Benign

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Topic: MIS-N: A Close Differential For Late-Onset Neonatal Sepsis

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Topic: A Rare Case of Multiple Abscess In A Newborn: Hyper IgE Syndrome

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AN UNUSUAL CAUSE OF SEVERE ECZEMA

Dr. Srinivas Jakka, Consultant in Paediatric Pulmonology and Allergy
Dr. Sagar Bhattad, Consultant in Paediatric immunology, and Rheumatology

CASE DETAILS

A 2 ½-year-old child male presented with rashes all over body from 1 year age. At the age of 1 year, he had acute gastroenteritis which was associated with papular rash on the legs. Between 12 to 18 months, he presented with recurrent erythematous rashes associated with itching and dermatographism. They were resolved with topical steroid creams. From 18 months onwards, the lesions increased in size and became less responsive to steroid creams. The entire body (except nose and ear lobes) including palms and soles were involved. Severe symptoms (itching and scratching) were leading to sleep disturbance every night for the child and the parents. The intense scratching resulted in bleeding and thickening of the skin. Sometimes purulent discharge was noted from the lesions on the scalp and skin during flareups. Mild temperature (99-100 F) was associated with flare-ups of rash; predominantly at nighttime. There was no involvement of the joints or mucous membranes or any other systemic involvement. His dietary intake was reduced during the flareups because of which his weight remained static at 8 kgs from the age of 1 year. He had received 3 to 4 courses of oral steroids over the course of time. His symptoms initially resolved after 3 to 4 days of oral steroids with recurrence of symptoms once the steroids were stopped. Gradually the rash and other symptoms became less responsive to oral steroids too. Investigations during the episodes revealed high white cell counts, eosinophil counts, and IgE levels. On examination, he had severe eczema all over the body. Some areas of the skin showed thickening and abscess formation. He also had cushingoid facies due to steroid usage. In view of severe eczema, he was started on Cyclosporin following which the skin symptoms improved. Further investigations including genetic tests revealed a pathogenic mutation in the DOCK 8 gene resulting in an autosomal recessive form of hyper IgE syndrome. The family was advised of bone marrow transplantation.

DISCUSSION

Severe eczema can be a marker of underlying immune defect. Early diagnosis of immunodeficiency is crucial for definitive management. Inborn errors of immunity such as Autosomal dominant hyper-IgE syndrome, DOCK 8 deficiency(AR-HIES), Immune dysregulation, polyendocrinopathy, and enteropathy,X-linked (IPEX) syndrome, Wiskott-Aldrich syndrome, Omenn syndrome, severe combined immune deficiency can have severe eczema. Awareness of associations between skin findings and immunodeficiency diseases may lead to early detection and treatment of serious immunologic defects.

TAKE HOME MESSAGE:
Severe Eczema can be the initial presenting feature of Inborn errors of immunity

Lehman H, Gordon C. The Skin as a Window into Primary Immune Deficiency Diseases: Atopic Dermatitis and Chronic Mucocutaneous Candidiasis. J Allergy Clin Immunol Pract. 2019 Mar;7(3):788-798. doi: 10.1016/j.jaip.2018.11.026. PMID: 30832893.

INFANTILE ONSET ARTHRITIS: NOT ALWAYS BENIGN

Dr. Anjani Gummadi, Consultant Paediatric Rheumatology and Immunology

CASE DETAILS

An 21 month old boy born to a non-consanguineous healthy parents presented with fever and multiple joint swellings. He was symptomatic since 7 months of age with recurrent knee swelling and fever at age of 7 and 14 months requiring analgesics and antibiotics, considering septic arthritis.

At 20 months of age, he developed bilateral knee swellings, associated with severe pain the morning and inability to talk. He also developed high grade fever for one month. Subsequently, multiple joints were involved and he was bed ridden.

Physical examination showed swollen and tender bilateral knee, ankle, wrist and elbow and splenomegaly. Possibilities of systemic juvenile idiopathic arthritis, RF positive polyarticular juvenile idiopathic arthritis, autoinflammatory syndrome were considered.

Laboratory findings revealed anemia (hemoglobin concentration 6.4 gm/dl), leukocytosis (leukocyte count 13,600/mm3), platelets of 9.68 lakh, elevated ESR (120 mm/hr) and CRP 48 mg/L, negative ANA and RF.

He was initiated on intravenous methylprednisolone pulse for 3 days after ruling out infection. He was discharged on tapering doses of oral prednisolone and subcutaneous methotrexate. At 2 months of follow up, child is afebrile, able to walk, arthritis and splenomegaly resolved.

DISCUSSION

Arthritis in children (including infants & neonates) can be due to infections, juvenile idiopathic arthritis, post infective, Kawasaki disease, MISC, connective tissue disorders, autoinflammatory syndromes, infiltrative etc. Pattern of joint involvement, associated systemic features provides the most important clues to type of arthritis[1].

Systemic juvenile idiopathic arthritis is a pediatric rheumatological disease characterized by the variable occurrence of chronic arthritis, high spiking fever, maculopapular rash during fever episodes, hepatomegaly &/or splenomegaly, lymphadenopathy, & serositis. Treatment includes the use of NSAIDS, corticosteroids, DMARDs or/and biological drugs[2].

It is important to diagnose and treat immediately to prevent complications such as macrophage activation syndrome (MAS), erosive arthritis, serosistis causing cardiac tamponade, interstitial lung disease and amyloidosis, etc. Chronic arthritis if left untreated can lead to growth impairment, limb length discrepancy & deformities.

TAKE HOME MESSAGE:
Joint swellings in infants need not always be Septic Arthritis. Pattern of joint involvement, associated systemic features provides the most important clues to type of arthritis.

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MIS-N: A CLOSE DIFFERENTIAL FOR LATE-ONSET NEONATAL SEPSIS.

Dr. Krishna Chaitanya, Consultant Paediatric Intensivist

INTRODUCTION

Multisystem inflammatory syndrome in newborns (MIS-N) is an entity that has been reported rarely by few centers throughout India. We present one such case which was managed successfully. MIS-N is an entity which manifest either following maternal COVID19 infection or postnatally acquired COVID -19 Infection. Neonatologists and pediatricians should be highly suspicious of the possibility of MISN which can result in multi-organ/system involvement if untreated.

CASE REPORT

A 28-day-old term boy baby born out of non-consanguineous marriage via LSCS presented to us with complaints of high grade fever and watery loose stools, excessive irritability for 2 days. Systemic examination was unremarkable. In view of excessive irritability, child was admitted and started on intravenous fluids. Investigations revealed a high C-reactive protein(CRP) and hence was given antimicrobials. Blood, CSF, and urine cultures were all negative. On day 3, CRP increased further following which the antibiotics were upgraded. The child had persistent symptoms despite adequate treatment and elevated CRP.

Clinical possibilities of late onset neonatal sepsis, MIS-N , inborn error of immunity were considered. In view of high inflammatory markers and sterile cultures, the possibility of MISN was higher. On probing, parents revealed that they both had febrile illness 1 week after delivery. Both the baby and mother were positive for Covid IgM and IgG antibodies. Inflammatory markers including ferritin, proBNP, and D dimer were elevated. 2D Echo was normal. Pediatric rheumatologist and immunologist's opinion taken, and baby was given IVIG 2g/kg following which the inflammatory markers came down, but fever persisted. Hence, intravenous Methylprednisolone pulses were administered following which the fever subsided by day 10 of illness and normal stool pattern was established. Baby was discharged home on oral prednisolone and LMWH and aspirin. By 6 weeks of follow up, drugs were tapered and stopped, inflammatory parameters normalised and 2D echo was normal.

DISCUSSION

MIS-C is a rare condition with an incidence of 316 per million. Neonates account for only 0.6% of MIS-C cases. MIS-N stands a close differential for LONS (late-onset Neonatal sepsis) in this era of the COVID pandemic. It is very important to differentiate from LONS, as the treatment modality is different and aggressive cardiac monitoring is required in MIS-N.

COVID IgM titres help in differentiating maternally acquired COVID antibodies from that of postnatal acquired COVID and must be checked after 2 weeks in the newborn. Also, a declining titer of IgG indicates a transplacental transfer whereas raising titers indicate a true infection in the baby.

In our index case, fever and GI involvement in the form of

diarrhea were predominant symptoms and mislead the treating pediatricians/neonatologist to treat as LONS. However, if there are no features of sepsis, both clinically and laboratory wise and no response to antibiotics, MIS-N needs to be considered as an important differential diagnoses.

TAKE HOME MESSAGE: Not all fevers in newborn are due to sepsis

	Day 1	Day 3	Day 7	Day 10	Day 14
Fever	Y	Y	Y	N	N
Gastroenteritis	Y	Y	N	N	N
CRP (mg/L)	10	40	56	8	4
Ferritin (ng/L)			926	789	654
D-dimer (mg/L)(N<0.5)			0.4	1.8	1.2
NT BNP			1270	664	345
2D Echo			N	N	
PCT (ng/dl) (N<0.5)		0.2	3.19	0.4	

Table 1.

Reference

1. Pawar, R.; Gavade, V.;Patil, N.; Mali, V.; Girwalkar, A.;Tarkasband, V.; Loya, S.; Chavan, A.; Nanivadekar, N.; Shinde, R.; et al. Neonatal Multisystem Inflammatory Syndrome (MIS-N) Associated with Prenatal Maternal SARS-CoV-2: A Case Series. Children 2021, 8, 572 . https://doi.org/10.3390/ children8070572.
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Our expert team for the case management:
Dr. Krishna Chaitanya (Ped. Intensivist),
Dr. Ramya Keerti (Ped. Intensivist),
Dr. Sunil Mohan (Neonatologist),
Dr. Anjani Gummadi (Pediatric Rheumatologist and Immunologist)

A RARE CASE OF MULTIPLE ABSCESS IN A NEWBORN: HYPER IgE SYNDROME

Dr. T.V Vijay Kumar, (Consultant Neonatologist & Paediatrician), | Dr. A V N Raja, (Consultant Neonatologist & Paediatrician)
Dr. S. Indu Sree, (Consultant Neonatologist & Paediatrician)

INTRODUCTION

Abscess in a newborn is a rare entity and could be due to an underlying inborn error of immunity. We present one such case with Hyper IgE syndrome.

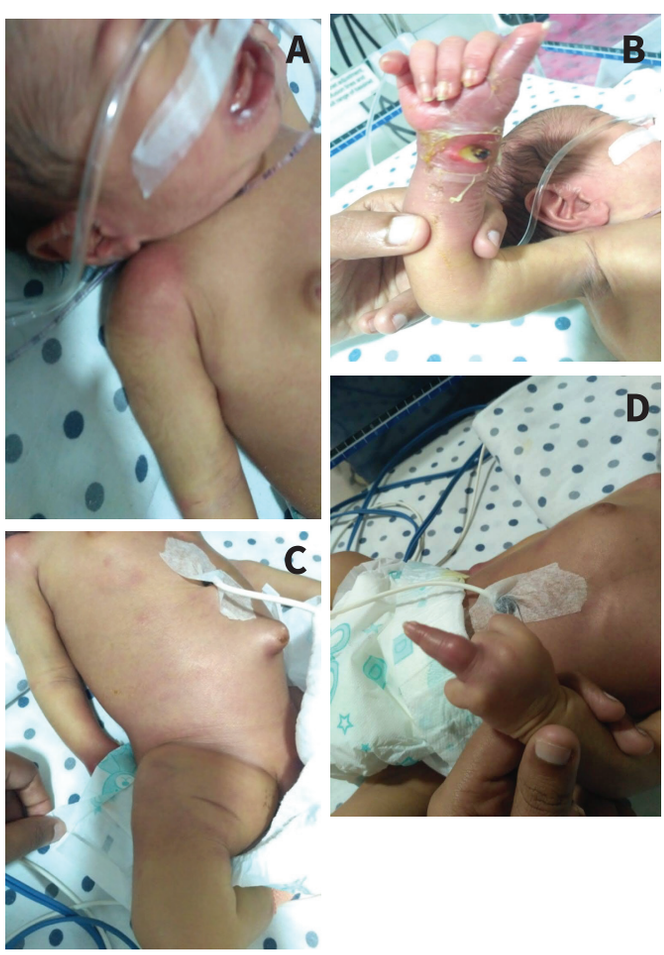
CASE REPORT

A 20-day old male baby presented to us with multiple swellings for 7 days. The swelling was initially noted on the right thigh, followed by right wrist, shoulder, and left ring finger.

On examination, he was active with stable vital signs. Mutiple abscess were noted (Fig.), rest of the systemic examination was normal.

His preliminary blood investigations showed thrombocytopenia and elevated CRP. His liver function tests showed elevated direct bilirubin, deranged AST and ALT. Ultrasound revealed abscess in right thigh with extension into the abdominal wall. He was started on antibiotics after obtaining blood cultures. Radionuclide scan showed no evidence of osteomyelitis. Incision and drainage of the abscess were done, intravenous antibiotics were continued. Blood culture and pus cultures showed the growth of MRSA (sensitive to Vancomycin). During the hospital stay he had right parotid abscess which was drained. He was given IV Antibiotics for 4 weeks and he was discharged on oral antibiotics followed by prophylaxis.

As he had multiple abscess possibility of inborn errors of immunity was considered. HIV serology of mother was non reactive. Lymphocyte count was normal for age. Nitroblue tetrazolium test was normal, hence ruled chronic granulomatous disease was ruled out. Immunoglobulin assay showed elevated Ig E levels >2500 IU/ml with normal levels of other immunoglobulins. Hence a provisional diagnosis of Hyper IgE was made and parents were
counselled for genetic analysis. At follow up baby was feeding well and gaining weight and lesions healed.



A. Abscess in right shoulder
B. Abscess in right wrist
C. Abscess in right ring finger
D. Abscess in right side of abdomen and thigh

Discussion

Hyperimmunoglobulin E syndrome is a rare primary immunodeficiency characterized by recurrent eczema, skin abscesses, lung infections, eosinophilia, and elevated levels of serum IgE . Two forms of hyper IgE syndrome (HIES) have been described including autosomal dominant (AD or type 1) and an autosomal recessive (AR or type 2).

The genetic defect seen in AD hyperIgE syndrome is in STAT3 which is a signal transducer and activator of transcription. These children are susceptible to recurrent bacterial and fungal infections . Clinical features will begin in first few days of life which consists of eczematous rash, multiple pyogenic abscess at various locations in the body , sinopulmonary infection . Other clinical features include dysmorphic facies , skeletal abnormalities, pneumatoceles, increased risk of fractures and risk of lymphoma[1-3]

Laboratory evaluation will show elevated IgE levels 1000 int.units/ml to 50,000 int.units/ml . Genetic diagnosis is confirmed by STAT3 mutation analysis . Management consists of drainage of abscess, prolonged antibiotics for 4-6 weeks and prevention of recurrent infections by appropriate antibiotic prophylaxus with Trimethoprim-sulfamethoxazole prophylaxis[4] , recombinant human interferon (IFN) gamma. Definitive treatment is by Hematopoetic stem cell transplantation.

TAKE HOME MESSAGE:
Severe infection in newborn warrant work up for inborn errors of immunity

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