

TEACHING FILE

GRAND ROUNDS

Ira Shah

YELLOW SKIN

Case:- A 9 years old girl presented with yellowish skin for 15 days without high colored urine. She also had fever, rash and yellow skin 3 months ago. She had neonatal hyperbilirubinemia at 1½ months of age. Mother had yellow skin 3 times in past 9 months. On examination, apart from yellow skin, there was no abnormality. Liver function tests were normal. Urine did not show any bile salts or bile pigments.

What is the diagnosis?

Expert's opinion:- Carotenemia. There was history of excessive carrot intake in the entire family. Ingestion of excessive amounts of carrots is the usual cause of carotenemia, but it can also be associated with ingestion of many other yellow vegetables, as well as some green vegetables which contain high levels of carotene (precursor of vitamin A). (1) As a precursor for vitamin A, carotene is converted in the body to retinol, which is one the most active forms of vitamin A. As a lipochrome, carotene contributes to the yellow component of normal skin. (2) Carotenemia is a benign condition; vitamin A poisoning does not occur despite massive doses of carotene because the conversion of carotene to vitamin A is slow. (1) Blood carotene levels are elevated. Carotenemia may be observed 4 to 7 weeks after initiation of a diet rich in carotenoids. (2) Hypothyroidism, diabetes mellitus, hepatic and renal diseases may be associated with carotenemia, but are not caused by ingestion of carotene. The absence of yellow pigment in the sclera and oral cavities distinguishes carotenemia from jaundice. A similar disorder, lycopenemia, is associated with an orange-yellow skin pigmentation as a result of ingestion of large amounts of tomatoes. (1) Therapy is based on dietary modification by reducing the carotene consumption. It may take several months before the skin color normalizes.

References

1. Lascari AD. Carotenemia. A review. Clin Pediatr (Phila). 1981; 20: 25-29.
2. Yao P. Carotenemia. Available at website: www.med.ucla.edu/modules/wfsectionperrticle.php?articleid=316. Accessed on 17th November 2014



DOI No.: 10.7199/ped.oncall.2015.13

WILM'S TUMOR

Case: - A 9 month old boy was referred for jaundice. He was diagnosed to have right sided Wilm's tumor at 6 months of age and underwent right sided nephrectomy. He was subsequently on chemotherapy of vincristine and daptomycin. But since past 1 month he had jaundice and chemotherapy had to be stopped. His bilirubin was 3.7 mg/dl (direct = 1.6 mg/dl), SGOT = 88 IU/L, SGPT = 42 IU/L, Total proteins = 7.1

gm/dl, albumin = 4.2 gm/dl, alkaline phosphatase = 604 IU/L, GGTP = 59 IU/L.

What is the likely cause of jaundice in this child?

Expert's opinion: Common causes of jaundice in a patient with Wilm's tumor are veno-occlusive disease (VOD), hepatotoxicity due to chemotherapy agents, obstruction of the inferior vena cava (IVC) due to tumor mass and infection related such as Hepatitis B and Hepatitis C.

VOD is a well-known complication in patients undergoing high-dose chemotherapy and bone marrow transplantation. The clinical signs of the disease are hepatomegaly, sudden weight gain with or without ascites, and jaundice. (1) Liver biopsy can confirm the clinical diagnosis of VOD by showing the small intrahepatic venules narrowed by an edematous concentric subendothelial zone containing fragmented red cells, debris, and fibrillar material; surrounding sinusoids are engorged and centrilobular hepatocytes are damaged. These changes lead to intrahepatic hypertension, hepatic enlargement, hyperbilirubinemia, peripheral edema, and ascites. (2) Ultrasound (USG) abdomen with doppler studies show reversal of flow in portal veins.

Hepatotoxicity may be observed during conventional chemotherapy. A syndrome characterized by ascites and hyperbilirubinemia has been reported following the administration of several antineoplastic drugs. These patients usually increased transaminases. (1) Both vincristine and actinomycin are known to cause severe hepatotoxicity. (3)

In our patient, USG abdomen showed recurrence of tumor and invasion of IVC that was causing the jaundice.

References

1. Bisogno G, de Kraker J, Weirich A, Masiero L, Ludwig R, Tournade M-F, Carli M. Veno-occlusive disease of the liver in children treated for Wilms tumor. Downloaded from UvA-DARE, the institutional repository of the University of Amsterdam (UvA). Available at website: [hdl.handle.net, 11245, 2.3285](http://hdl.handle.net/11245/2.3285). Accessed on 23rd December 2014
2. Mc Donald GB: Veno-occlusive disease of the liver following marrow transplantation. Marrow Transpl Rev. 1993; 3:49-56
3. McVeagh P, Ekert H. Hepatotoxicity of chemotherapy following nephrectomy and radiation therapy for right-sided Wilms tumor. J Pediatr. 1975; 87:627-8



DOI No.: 10.7199/ped.oncall.2015.12

CUTANEOUS TUBERCULOSIS

Case: - A 3 years old boy presented in December 2014 with reddish patch of 1 cm diameter over left cheek for the past 1½ years. He had been to various

dermatologists for the same but had no relief. A biopsy done in October 2014 showed patchy nodular tuberculoid granulomatous infiltrate made up of lymphocytes, plasma cells, histiocytes and epithelioid cells with occasional langerhans giant cells suggestive of tuberculosis. There was no contact with a patient having TB. He was started on antituberculous therapy (ATT) and referred for further management. On presentation, weight was 12.7 kg, height was 90 cm. He had chancre over left cheek. Other systems were normal. On enquiry, mother gave history of taking bath from bore well water prior to onset of rash. His mantoux test was 1.7 cm and chest X-Ray was normal. He was continued on ATT and clarithromycin was added to cover atypical organisms.

How to manage cutaneous TB?

Expert's opinion: Cutaneous tuberculosis (TB) is skin TB that can be caused by *Mycobacterium tuberculosis* (MTB), *Mycobacterium bovis*, non-tuberculous mycobacteria (NTM) and the Bacille Calmette-Guérin (BCG) vaccine. (1,2) Nearly every pathogenic species of NTM may cause skin and soft tissue infections. It is quite rare and difficult to diagnose. TB is an airborne transmissible disease with skin manifestations presenting as a result of hematogenous spread or direct extension from a latent or active foci of infection. However, primary inoculation may occur as a direct introduction of the mycobacterium into the skin or mucosa of a susceptible individual by trauma or injury with water or other contaminated products (1,3) Through direct inoculation, it can cause tuberculous chancre, tuberculosis verrucosa cutis and lupus vulgaris. Through hematogenous spread, it can cause acute miliary TB, metastatic TB abscess (gummatous TB), papulonecrotic tuberculid and lupus vulgaris. Spread from contiguous area can cause scrofuloderma, orificial TB. (4) Tuberculous chancre typically follows a penetrating injury that results in the direct introduction of mycobacterium into the skin or mucosa of an individual with no previous TB infection. Within 2 to 4 weeks, an inflammatory papule develops at the inoculation site and evolves into a firm,

shallow, non-tender, non-healing, undermined ulcer with a granulomatous base (1) Diagnosis is based on histopathology, culture from the skin lesion. However yield is poor and PCR may be a more useful test which also helps in identification of the species. Screening for internal organ assessment should be done to rule out endogenous TB. The treatment of cutaneous TB is the same as that for systemic TB. NTM are resistant to conventional antituberculous drugs and macrolides form the mainstay of therapy. In our patient, we were unable to do PCR test on the chancre. In view of exposure to bore well water, and isolated cutaneous TB, he was suspected to have inoculation as direct exposure and thus cover for NTM was also added.

References

1. Frankel A, Penrose C, Emer J. Cutaneous Tuberculosis. J Clin Aesthet Dermatol. Oct 2009; 2: 19-27.
2. Wentworth AB, Drage LA, Wengenack NL, Wilson JW, Lohse CM. Increased incidence of cutaneous nontuberculous mycobacterial infection, 1980 to 2009: a population-based study.
3. Alcaide F, Esteban J. Cutaneous and soft skin infections due to non-tuberculous mycobacteria. Enferm Infecc Microbiol Clin. 2010;28 Suppl 1:46-50
4. Barbagallo J, Tager P, Ingleton R, Hirsch RJ, Weinberg JM. Cutaneous tuberculosis: diagnosis and treatment. Am J Clin Dermatol. 2002; 3:319-328. Mayo Clin Proc. 2013; 88:38-45

DOI No. : 10.7199/ped.oncall.2015.14

From: Medical Sciences Department, Pediatric Oncall, Mumbai.

Address for Correspondence Dr Ira Shah, 1/B Saguna, 271, B St Francis Road, Vile Parle (W), Mumbai 400056.

