Acrocyanosis with intrahepatic carcinoid tumor

Sir,

Paraneoplastic disorders occur due to substances released from the internal tumors or immune cross-reactivity with host tissue. Common mucocutaneous paraneoplastic disorders include acanthosis nigricans, dermatomyositis, pyoderma gangrenosum, and erythroderma.[1] Acrocyanosis means symmetrical, painless bluish discoloration of the distal parts of the body. It is classified as primary, with no apparent underlying cause, and secondary, when associated with underlying pathology.[2]

A 49-year-old man presented with discoloration of the fingers and toes of one month duration. He denied pain and swelling of the extremities and aggravation of symptoms in cold climate. He gave a history of intermittent abdominal pain over the right upper quadrant region associated with weight loss for the past one

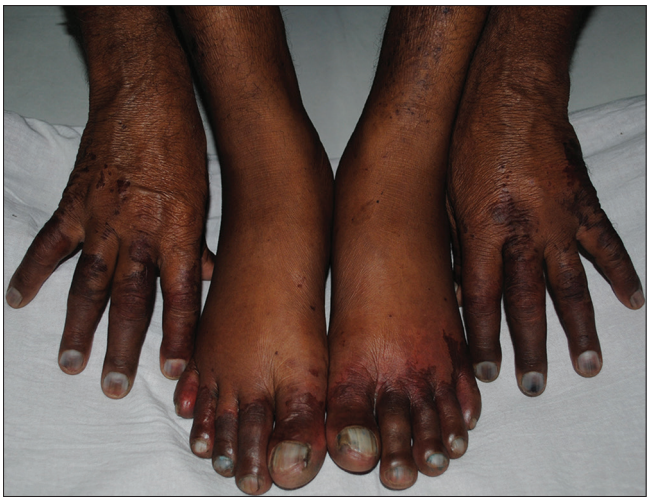

Figure 1: Clinical photograph showing acrocyanosis

year and denied any history of diarrhea or flushing episodes. Examination revealed low body mass index (19.6 kg/m^2), acrocyanosis [Figure 1], and tender hepatomegaly. Rest of the systemic examination was essentially normal. Ultrasonography and computed tomography scan of the abdomen showed a heterogeneous solid lesion of 8 × 8 cm in the right lobe of the liver with occasional areas of breakdown [Figure 2]. The lesion showed enhancement in the arterial phase with delayed washout in the portal phase. His hematological, biochemical parameters and gastrointestinal endoscopy were normal. Serum chromogranin A level was elevated (225 ng/mL) and other tumor markers, such as Alpha-fetoprotein (AFP) and Carcinoembryonic antigen (CEA), were normal. Liver biopsy revealed tumor cells arranged in a trabecular nest of uniform cells separated by delicate vascular stroma and stained positive with chromogranin and synaptophysin. Positron emission tomography scan or octreotide scan was not done due to nonavailability. The patient was planned for lobectomy of the liver or chemoembolization but succumbed due to complicated nosocomial pneumonia.

Acrocyanosis as the presentation of carcinoid syndrome has been reported only once previously.[3] Acrocyanosis is described as Flying Dutchman, and is frequently associated with the palmoplantar hyperhidrosis. The differential diagnoses include Raynaud's phenomenon and pernio- and erythromelalgia. The differentiation with Raynaud's phenomenon is mainly clinical and acrocyanosis is characterized by prolonged bluish discoloration, symmetry, and lack of paroxysmal pallor. Nailfold capillaroscopy identifies the capillary flow patterns and also helps in the diagnosis in difficult cases. Primary acrocyanosis is a disease seen in 2nd to 3rd decade and secondary acrocyanosis is seen due to connective tissue disorders, hematological diseases, neoplasms, infections, drugs, and eating disorders. The treatment of primary acrocyanosis is controversial and

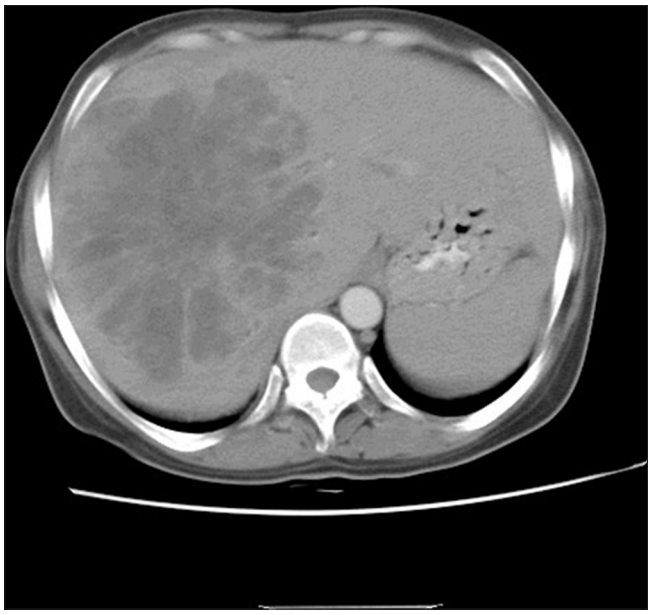

Figure 2: Computed tomography scan of the abdomen showing heterogeneous mass lesion in the liver

includes nicotinic acid derivatives, beta-blockers, and low molecular weight dextran.

Carcinoid tumors are well-differentiated tumors originating from the diffuse endocrine system outside the pancreas and thyroid. The syndrome develops when vasoactive substances produced by the tumor enter the systemic circulation without undergoing metabolic degradation. Paraneoplastic acral vascular syndromes are reported rarely. The common presentations include peripheral gangrene, acrocyanosis, and Raynaud's phenomenon.[4] The presentation is more common in adult males beyond the fifth decade. The most common malignancies associated are adenocarcinomas and metastatic deposits. Acral vascular syndromes regress in majority with definitive treatment, and prostacyclin infusions are given in resistant cases. Acrocyanosis as a paraneoplastic feature suggests a mortality rate of more than 50% in 2 years.

K.V.S. Hari Kumar, Amit Kumar, Deepak Tomar, A.K. Gupta[1]

Department of Endocrinology and [1]Oncology, Command Hospital, Lucknow, Uttar Pradesh, India

Address for correspondence:
Dr. K.V.S. Hari Kumar,
Department of Endocrinology, Command Hospital,
Lucknow - 226 002, Uttar Pradesh, India.
E-mail: hariendo@rediffmail.com

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