

McCune-Albright syndrome

Suparat Thamrat D.D.S., Grad. Dip. (Oral Medicine)¹

Pannipa Thumasombut D.D.S., Grad. Dip. (Oral Medicine)¹

Samornroj Arporniem D.D.S., Grad. Dip. (Oral Medicine)¹

Wichitsak Cholitgul B.Sc., D.D.S., Diplomate, Thai Board of Oral Diagnostic Science²

¹Graduate student, Department of Oral Medicine, Faculty of Dentistry, Chulalongkorn University

²Department of Radiology, Faculty of Dentistry, Chulalongkorn University

Abstract

This literature review aims to report various defects appeared in patients with McCune-Albright Syndrome which is a rare, multisystem disorder caused by GNAS 1 gene mutation. This gene code for the stimulatory subunit of G protein involved in cyclic adenosine monophosphate synthesis, resulting in persistently elevated cyclic adenosine monophosphate levels. The clinical manifestation of the disorder is characterized by the triad of café-au-lait cutaneous spot, polyostotic fibrous dysplasia and endocrinopathy. Fibrous dysplasia frequently affects the craniofacial bones and dental tissue causing disfigurement and developmental anomalies of teeth. These symptoms can be early detected by the first sign of fibrous dysplasia during an oral examination and leading to appropriate diagnosis of McCune-Albright Syndrome. The diagnosis usually relies on clinical informations as facial asymmetry including radiographic features as ground glass appearance. There is no specific treatment for this syndrome, usually the symptomatic treatment with long term medical follow-up for the endocrine disturbances is recommended.

(CU Dent J. 2014;37:113-22)

Key words: *café-au-lait cutaneous spot; McCune-Albright Syndrome; polyostotic fibrous dysplasia*

Correspondence to Suparat Thamrat, Suparat6516@yahoo.com