LETTER TO THE EDITOR

Systemic abnormalities associated with cutis marmorata telangiectatica congenita

To the Editor—The article by Soo et al[1] is highly interesting. Cutis marmorata telangiectatica congenita assumes clinical significance as it may be associated with a number of congenital systemic abnormalities besides those mentioned by Soo et al.

Nearly 68% of patients with cutis marmorata have a congenital abnormality, the most common being body asymmetry. Cutis marmorata has also been shown to be associated with the chiari malformations, macrocephaly, hemimegalencephaly, bilateral cortical dysplasia, calvarial haemangioma and cavum septi pellucidum cysts.[2] In fact, this subset of patients may have developmental delay and are now classified under a separate subset: macrocephaly-cutis marmorata telangiectatica congenita (M-CMTC). Similarly, ocular abnormalities such as late-onset paediatric glaucoma have been reported in some patients.[3] Cardiac defects such as atrial septal defects and pulmonary vein stenosis may also occur. Haematologic abnormalities such as myelodysplasia and even gastro-intestinal abnormalities such as imperforate anus may occur.[4] Hypospadias is another common abnormality seen in association with cutis marmorata. Overall, the prognosis in cutis marmorata is very good, as almost all patients show an improvement in the cutaneous lesions during infancy. Rarely, serious complications such as bleeding episodes, gangrenous ulceration and hypovolaemic shock may occur.[5] The disease may occasionally be fatal, especially in patients with M-CMTC.[2]

Physicians need to be aware of these associated systemic defects in order to decrease the morbidity and mortality associated with cutis marmorata.

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References

1. Soo MT, Lo KK, Leung LC. Cutis marmorata telangiectatica congenita. Hong Kong Med J 2007;13:491-2.
2. Akcar N, Adapinar B, Dinleyici C, Durak B, Ozkan IR. A case of macrocephaly-cutis marmorata telangiectatica congenita and review of neuroradiologic features. Ann Genet 2004;47:261-5.
3. Murphy CC, Khong CH, Ward WJ, Morgan WH. Late-onset pediatric glaucoma associated with cutis marmorata telangiectatica congenita managed with Molteno implant surgery: case report and review of the literature. J AAPOS 2007;11:519-21.
4. Khanna N, Curry ML, Carder KR. Cutis marmorata telangiectatica congenita in unique association with imperforate anus and myelodysplasia. J Am Acad Dermatol 2005;53:904-5.
5. Hu IJ, Chen MT, Tai HC, Tsao PN, Chou HC, Hsieh WS. Cutis marmorata telangiectatica congenita with gangrenous ulceration and hypovolaemic shock. Eur J Pediatr 2005;164:411-3.