Griscelli Syndrome with Fibronodular Sclerodermatous Chronic Graft Versus Host Disease

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A 4-year male presented to us with Griscelli syndrome with Rab 27A mutation. His elder sibling died of same illness at 8 years' age. He received allogeneic matched sibling donor peripheral blood stem cell transplant (PBSCT) from his sister in October 2015. He received conditioning regime with busulfan, cyclophosphamide and horse antithymocyte globulin [1]. Tacrolimus and methotrexate were used as graft versus host disease (GVHD) prophylaxis. He engrafted on day 13 of PBSCT.

He developed steroid refractory GVHD of skin, liver and gut which was treated with basiliximab and mycophenolate mofetil. Gradually his diarrhea responded

and liver functions started to normalize. Subsequently he developed CMV reactivation which was treated with ganciclovir. His skin showed fibronodular lesions on follow-up (Fig. 1a). At 1 year follow-up, he is doing well with 99% donor chimerism. He is on tapering doses of mycophenolate at present. His hair are still silver-gray (Fig. 1b). He has mosaic pattern of skin and still requires moisturizing agents for dry skin.

Chronic GVHD is known complication of bone marrow transplant. Cutaneous GVHD is usually diagnosed clinically [2]. Various treatment option available include corticosteroids, topical therapy and maintaining integrity of

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Fig. 1 Photograph showing **a** fibronodular lesions of GVHD and **b** Mosaic pattern of skin due to GVHD

skin [3]. This child has steroid refractory acute GVHD which went into remission and later evolved into chronic GVHD of skin.

Compliance with ethical standards

Conflict of interest Rahul Naithani declares that he has no conflict of interest.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent Informed consent was taken from mother.

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