Authors' reply

Sir,

We thank the authors for their interest in our letter "Recurrent spontaneous forehead ecchymoses with headache: A distinctly curious phenomenon."[1] The authors suggest that spontaneous extracranial hemorrhagic phenomena could be a form of Gardner-Diamond syndrome, and that anticardiolipin antibodies should be tested in these patients.

Both these conditions are poorly understood entities, affecting predominantly women and characterized by spontaneous bruising in the absence of hematological or clotting abnormalities. However, there are certain important differences between the two. Spontaneous extracranial hemorrhagic phenomena is characterized by recurrent ecchymoses on the face, primarily the forehead and periorbital areas, triggered by episodes of headache. The ecchymotic patches usually occur on the same side as the headache, but may occur on the contralateral side as well.[2] Ecchymoses in Gardner-Diamond syndrome, on the other hand, usually occur on the lower and upper extremities (44–67%), with facial involvement (22%) being less common. Patients typically have a background psychological disorder, and common triggers include stress, physical work, minor trauma or surgery, or may occur spontaneously as well. Sometimes, bruising episodes may be accompanied by general symptoms such as fever, arthralgias, myalgias, headache and dizziness.[3,4] Further, ecchymoses in Gardner-Diamond syndrome are usually painful, in contrast to spontaneous extracranial hemorrhagic phenomena where they are described as asymptomatic.

The association of headache with Gardner-Diamond syndrome is not as strong as is with spontaneous extracranial hemorrhagic phenomena. Headache in Gardner-Diamond syndrome, may occur as a part of prodromal symptoms instead of being a specific trigger.[3,4] Spontaneous extracranial hemorrhagic phenomena occurs in association with a primary headache disorder, usually migraine or trigeminal autonomic cephalalgia, whereas such a characterization of headache has not been done in Gardner-Diamond syndrome.[2]

Further, the proposed hypotheses regarding the pathogenesis of spontaneous extracranial hemorrhagic phenomena and Gardner-Diamond syndrome are also different. Spontaneous extracranial hemorrhagic phenomena is hypothesized to occur as a result of trigeminal-autonomic reflex activation (as part of the primary headache disorder) leading to increased parasympathetic outflow to head and neck vasculature,[2] while autosensitization to one's erythrocytes and psychological stress is postulated to play a role in the causation of Gardner-Diamond syndrome.[3,4] Given the dissimilarities in the location, symptoms and triggers of ecchymoses as well as their pathophysiology, it seems unlikely that spontaneous extracranial hemorrhagic phenomena and Gardner-Diamond syndrome are related disorders.

To our knowledge, anticardiolipin antibodies have not been tested in patients with spontaneous extracranial hemorrhagic phenomena. The utility of testing anticardiolipin antibodies in patients with Gardner-Diamond syndrome is also not well established, and needs further evaluation. We came across only one report of Gardner-Diamond syndrome associated with anticardiolipin antibodies in the literature.[5] It should be noted that anticardiolipin antibodies are relatively nonspecific, being positive in a variety of infections, leukemias, solid-organ malignancies and even healthy individuals.[6] Intradermal auto-erythrocyte sensitization test is considered a more reliable diagnostic test for Gardner-Diamond syndrome and maybe a better method if one wishes to test its association with spontaneous extracranial hemorrhagic phenomena.[4]

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Conflicts of interest
There are no conflicts of interest.

Vishal Gupta, Snehal Agrawal, G. N. Abhishek, Shipra Agarwal[1], Kanika Sahni

Departments of Dermatology and Venereology and [1]Pathology, All India Institute of Medical Sciences, New Delhi, India

Corresponding author:
Dr. Kanika Sahni,
Department of Dermatology and Venereology, All India Institute of Medical Sciences, Ansari Nagar, New Delhi - 110 029, India.
kanu.sah@gmail.com

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