Kaposi's Sarcoma Restricted to an Immunocompromised District

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We read with interest, albeit a lack of surprise, the case of Kostaki et al. [1]. In particular, the authors described the onset of KS nodules solely on the right palm of a 63-year-old man who had undergone repeated and minimally invasive surgical procedures (needle fasciotomy) for Dupuytren’s disease. No lesions were present elsewhere, and the patient had no signs of systemic immunodeficiency.

The case is of interest because it seems to be the first report of an immunocompetent patient who has developed a KS after a localized trauma. However, our lack of surprise originates in the common knowledge that, even in the absence of systemic immunodeficiency, numerous and varied immunity-related disorders (infections, tumors, immune reactions) can appear on and remain limited to body regions that have been immunologically destabilized by chronic lymph stasis, herpetic infections, ionizing radiation, burn, or trauma (even the minimal trauma occurring with vaccinations) [2]. In their report, Kostaki et al. [1], by noticing that KS developed after repeated surgeries in a unique peculiar localized area with a dense lymphatic network, put forward the hypothesis that tissue injuries involving the lymphatic routes could play a central role in the occurrence of KS [1]. A large body of evidence that has been accumulated over the last three decades [3–10] strongly supports this concept, which is much more than a hypothesis. In fact, any persistent hindrance to lymph flow in a given body area impairs local immune surveillance (by disrupting trafficking of the immunocompetent cells) and stimulates vicarious angiogenesis (by promoting development of a collateral lymphatic or blood vascular network in the involved district). When the local immune control begins to fail, the district with lymph flow dysfunction becomes an immunologically vulnerable area, predisposed to malignancies – chiefly those of vascular origin, such as KS or other angiosarcomas (Stewart-Treves syndrome) – because of the continual angiogenic stimulus [6, 7, 11]. Put simply, lymph stasis results in immune stasis, which in turn may result in opportunistic oncogenesis, especially in reference to vascular tumors.

From this lymphological perspective, it has even been assumed that KS and Stewart-Treves angiosarcoma may not be completely different entities, but merely variant expressions of a similar underlying abnormality consisting of impairments in lymph drainage and immune control [6]. This is supported by the existence of borderline cases [12]. Clinical observations and experimental investigations have confirmed the parallel course of the lymphatic and immune functions in body regions affected with KS [3–9] or Stewart-Treves angiosarcoma [13, 14]. In this light, the two conditions may only differ in etiology, with human herpes virus 8 (HHV-8) always being causative of KS, while unknown viruses might be responsible for other angiosarcomas (though occasionally the same HHV-8 has been suspected) [12, 15]. The transformation of a common wart into a squamous cell carcinoma in a patient with chronic lymphedema [16] clearly indicates how a viral infection (human papilloma virus infection in this example) can evolve into an ‘opportunist’ tumor (squamous cell carcinoma in this example) in the presence of persistent lymph stasis.

In summary, the case reported by Kostaki et al. [1] features a typical example of an immunocompromised district due to repeated surgical procedures that impaired both lymph circulation and immune control in the traumatized area. Bearing in mind that an immunocompromised district may harbor opportunist and immunity-related disorders – such as infections, tumors and immune reactions [2] – the onset of KS lesions is not at all surprising here, since KS has an infectious etiology (HHV-8), a tumor morphology (angiogenic neoplasm), and an immune pathogenesis (immunosuppression often confined to acral body regions [3–10]).

Disclosure Statement
The authors declare no conflict of interest.

References


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