Cells to Surgery Quiz: January 2020

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WHAT IS YOUR DIAGNOSIS?

Figure 1. Images courtesy of Toby Maurer, MD, University of California, San Francisco.

Editorial note: Welcome to the Journal of Investigative Dermatology (JID) Cells to Surgery Quiz. In this monthly online-only quiz, the first question (“What is your diagnosis?”) relates to the clinical image shown, while additional questions concern the findings reported in the JID article by Chattopadhyay et al. (2019) (https://doi.org/10.1016/j.jid.2019.04.031).

Detailed answers and a list of relevant references are available following the quiz questions below.

QUIZ QUESTIONS

1. A man who is HIV-positive presents with the following painless skin lesions in addition to similar lesions on the face and hard palate. He denies taking any medications. What is your diagnosis?

   a. Eruptive keloids
   b. Kaposi sarcoma
   c. Pityriasis rosea
   d. Bacillary angiomatosis
   e. Angiosarcoma

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2. What would be first-line therapy for this patient?
   a. Intralesional vinblastine
   b. Mohs micrographic surgery
   c. Erythromycin
   d. Cryotherapy
   e. Highly active antiretroviral therapy

3. Based on the findings by Chattopadhyay et al. (2019), this patient would be at highest risk for developing a secondary primary cancer of which type?
   a. Hodgkin lymphoma
   b. Stomach cancer
   c. Breast cancer
   d. Prostate cancer
   e. Malignant melanoma

See following pages for detailed answers.
DETAILED ANSWERS

1. A man who is HIV-positive presents with the following painless skin lesions in addition to similar lesions on the face and hard palate. He denies taking any medications. What is your diagnosis?

CORRECT ANSWER: b. Kaposi sarcoma

Multiple painless violaceous plaques and nodules on the trunk, oral mucosa, and face in the setting of HIV-positivity are highly suggestive of Kaposi sarcoma (KS). KS is an angioproliferative spindle cell tumor of endothelial cells infected with human herpesvirus-8. It is divided into four clinical subtypes: classic, endemic or African, immunosuppression-related, and AIDS-associated (Mui et al., 2019). KS is the most common tumor arising in patients that are HIV-positive, and CD4 cell count is inversely correlated with its incidence (Lodi et al., 2010). Although AIDS-associated KS is often more aggressive than the classic form, the widespread use of highly active antiretroviral therapy has led to a decrease in incidence (Curtiss et al., 2016).

Discussion of incorrect answers

a. Eruptive keloids: Eruptive keloids have been described in association with systemic disease such as scleroderma, nephrogenic systemic fibrosis, and letrozole therapy. Rather than the violaceous plaques and papules of the KS picture here, eruptive keloids present with multiple skin-colored to hyperpigmented, firm, claw-like plaques (Meade et al., 2015).

c. Pityriasis rosea: Although the lesions of both pityriasis rosea (PR) and KS may follow skin tension lines, the exanthem of PR classically presents with the herald patch, a thin, oval, scaly plaque on the trunk, followed by multiple smaller pink, thin, oval papules and plaques with a fine collarette of scale. Although the cause of PR has not been fully elucidated, some authors have suggested a connection with human herpesviruses 6 and 7 (Eisman and Sinclair, 2015).

d. Bacillary angiomatosis: Bacillary angiomatosis is an infectious disease caused by organisms of the genus Bartonella, leading to vascular proliferation (Kaçar et al., 2010). The lesions of bacillary angiomatosis may mimic KS and may ulcerate and crust. The Warthin-Starry stain may be used to identify the bacilli on histology (Zapata et al., 2013). Erythromycin or doxycycline is the treatment of choice (Stevens et al., 2014).

e. Angiosarcoma: Angiosarcoma is an aggressive malignancy of vascular endothelial cells that most commonly presents as a red-purple purpuric plaque or nodule on the head and neck region of an elderly person. Radiation-associated angiosarcoma can be seen as a complication of radiation therapy for breast carcinoma (or other malignancies), with a median latency period of five years. Development of angiosarcoma in the setting of chronic lymphedema has also been described. Surgery with wide margins in combination with radiation therapy is the primary treatment, and the prognosis often remains poor (Shustef et al., 2017).

2. What would be first-line therapy for this patient?

CORRECT ANSWER: e. Highly active antiretroviral therapy

Highly active antiretroviral therapy (HAART) is first-line therapy for AIDS-associated Kaposi sarcoma (KS) (Dilorenzo et al., 2007). Although HAART therapy has decreased the incidence and severity of AIDS-associated KS, the phenomenon of immune reconstitution inflammatory syndrome can cause worsening of symptoms and progression of disease. Close follow-up is recommended (Letang et al., 2013). Categorizing patients using extent of tumor, immune status, and severity of systemic illness can help to guide additional therapeutic choices (Krown et al., 1989). Localized treatments are often a useful adjunct to systemic therapy for symptomatic or cosmetically bothersome lesions. Localized treatments include surgical excision, cryotherapy, radiation therapy, laser surgery, photodynamic therapy, and intralesional chemotherapy such as vinblastine or vincristine (Soleymani and Bennett, 2019). See the incorrect answers for further discussion of select localized treatments.

Discussion of incorrect answers

a. Intralesional vinblastine: Vinblastine is a vinca alkaloid that disrupts microtubular function (Brambilla et al., 2010). Intralesional vinblastine can be injected into individual KS lesions to induce regression, though it does not prevent new lesions (McCormick, 1996). Intralesional vinblastine has been effective for oral lesions of AIDS-associated KS as well as some cutaneous lesions (Boudreaux et al., 1993). Intralesional therapies are most often limited to a small number of symptomatic lesions or those in cosmetically sensitive areas. A similar medication, vincristine, has been reported as successful for intralesional injection in patients with classic KS (Brambilla et al., 2010).

b. Mohs micrographic surgery: Mohs micrographic surgery (MMS) has been reported for successful removal of symptomatic lesions on the feet and lower extremities in an elderly patient with classic KS.
**Discussion of incorrect answers**

b. **Stomach cancer**: Neither the RR of stomach cancer after KS nor the RR of KS after stomach cancer was increased. Mild bidirectional RR of stomach cancer and invasive skin squamous cell carcinoma (SCC) was found, with a risk of stomach cancer after invasive SCC of 1.56 and a risk of invasive SCC after stomach cancer of 1.19 (Chattopadhyay et al., 2019).

c. **Breast cancer**: Neither the RR of breast cancer after KS nor the RR of KS after breast cancer was increased. Mild bidirectional RR of breast cancer and invasive SCC was found, with a risk of breast cancer after invasive SCC of 1.35 and a risk of invasive SCC after breast cancer of 1.65 (Chattopadhyay et al., 2019).

d. **Prostate cancer**: Neither the RR of prostate cancer after KS nor the RR of KS after prostate cancer was increased. Mild bidirectional RR of prostate cancer and invasive SCC was found, with a risk of prostate cancer after invasive SCC of 1.20 and a risk of invasive SCC after prostate cancer of 1.53. Additionally, the risk of developing Merkel cell carcinoma (MCC) after prostate cancer was 1.92 (Chattopadhyay et al., 2019).

e. **Malignant melanoma**: Neither the RR of malignant melanoma after KS nor the RR of KS after malignant melanoma was increased. Bidirectional RR of melanoma and invasive SCC was found, with a risk of melanoma after invasive SCC of 3.81 and a risk of invasive SCC after melanoma of 3.89. Additionally, bidirectional RR of melanoma and MCC was found, with a risk of melanoma after MCC of 6.05 and a risk of MCC after melanoma of 3.30 (Chattopadhyay et al., 2019).

**REFERENCES**


