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Radiological-Pathological Correlation

Sclerosing Kaposi's sarcoma of the adrenal gland in an HIV-infected patient under antiretroviral therapy☆

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1. Introduction

Visceral Kaposi's sarcoma(KS) is relatively uncommon in the absence of mucocutaneous lesions, andadrenal gland involvement is remarkably rare. Since the introduction of combined antiretroviral therapy (ART) the incidence of KS, and especially that with visceral involvement, has declined markedly in HIV-infected patients [1]. Kaposi sarcoma appearing under ART seems to be less aggressive, but there are not reported differences in its histopathologic features. Although regression of the neoplasmis not uncommon, histologic changes related to response to the treatment are poorly described, particularly in visceral forms [2]. We report a case of an apparently primary adrenal Kaposi's sarcoma with a peculiar sclerosing pattern in an HIV seropositive patient who was under ART.

2. Case presentation

A 53-year-old male was diagnosed in 2005 with HIV infection, beginning ART 4 years after the diagnosis. Since then viral loads remained under control only with some unremarkable blips, CD4 counts ranging from 244 to 489 cells/mm^3. In 2015 a thoraco-abdominal computerized tomography (CT) performed after an episode of dyspnea found a right renal tumor of 96 mm, as well as a low-suspicion pulmonary nodular lesion of 11 mm, and a splenic hemangioma of 30 mm. A partial nephrectomy was performed, demonstrating a papillary renal cell carcinoma type I with negative margins. After two years of follow-up,a right adrenal nodular lesion of 9 mm was found, that displayed progressive contrast uptake from the periphery on CT, and showed slight growth in a subsequent magnetic resonance imaging (MRI), prompting adrenalectomy with the clinical suspicion of metastasis. Grossly, a multinodular lesion with red and white merged areas partially replaced the adrenal grand (Fig. 1). Histologically it consisted of a proliferation of small vessels with solid spindle cell areas. Extensive sclerotic areas with lower density of vessels and scattered neoplastic cells were observed. On immunohistochemistry neoplastic cells were diffusely positive for endothelial markers (CD31 and CD34) and human herpes virus 8 (HHV-8) (Fig. 2). A new complete physical examination excluded mucocutaneous involvement. Retrospective examination of CT images revealed that the lesion was apparent only since 2017, with the patient being under ART and with CD4 count > 200 cells/mm^3. Both the pulmonary nodule and the splenic hemangioma remained unchanged since the first detection. Therefore, a wait and see approach was decided with a control CT and MRI imaging 6 months after the surgery, which it showed the patient was free of disease.

3. Discussion

The present case is an isolated HIV-related visceral KS appearing during ART. Visceral involvement in the absence of mucocutaneous involvement is rare, even more if CD4 count is upon ≥200 cells/mm^3, although patients developing HIV-related KS despite apparent correction of their immunodeficiency have been reported [3].

A striking peculiarity in our case was the presence of extensive sclerotic areas, a feature not reported previously in visceral KS. Several variants of cutaneous KS have been described, including a keloidal form [4,5]. Regressed skin lesions may also become sclerotic, especially those that are subjected to intralesional chemotherapy injections [4]. A single report on adrenal involvement described fibrosis after systemic chemotherapy [2]. In our patient, the tumor appeared under ART and he didn't receive chemotherapy for its KS. Even so, sclerosis could be interpreted as spontaneous regression. Clinical regression is a well-known phenomenon both in cutaneous and visceral KS in response to therapy, but there are few reports on histologic features of regression and they are limited to cutaneous lesions [6]: regressed lesions are defined by clinical reduction in size and show diminished or absent proliferation of spindle cells. In most cases, therapy related regression is associated with a loss of HHV8 expression in neoplastic cells [7]. In our case, according to imaging, the lesion was still growing, and HHV8

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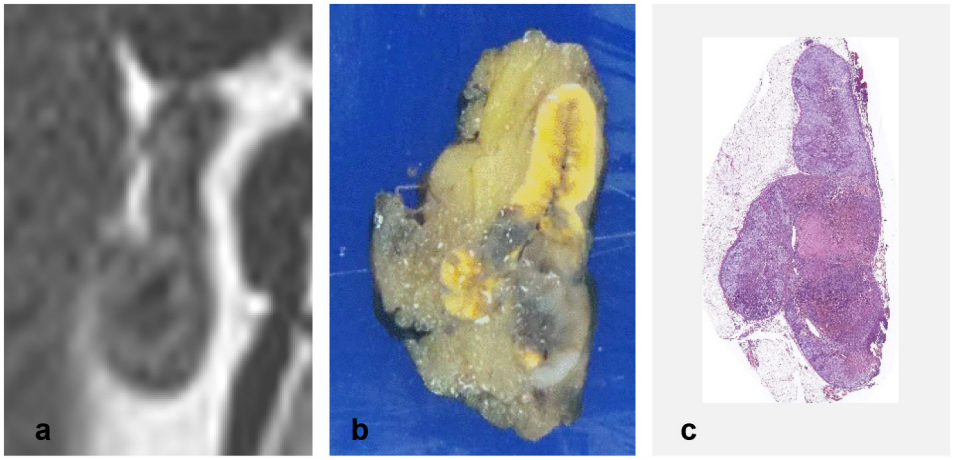

Fig. 1. a) MRI showing the adrenal lesion b) Gross and c) microscopic appearance of the KS in a whole adrenal section.

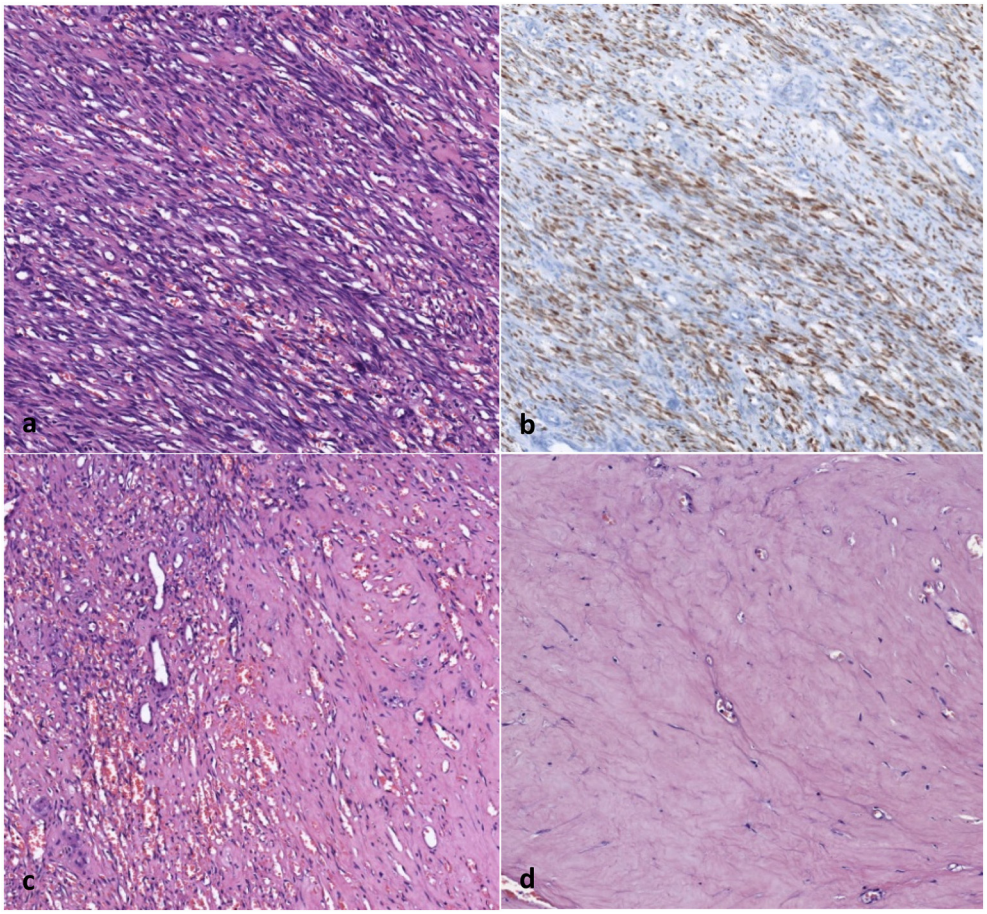

Fig. 2. a) Microscopical appearance of the KS in a solid area. b) Positive HHV8 immunostaining. c) Transition between an active and a sclerosing area of the lesion. d) Sclerosing area of the tumor.

remained strongly positive indicating that this case would represent a previously undescribed sclerosing variant of visceral KS, comparable to the keloidal counterpart in the skin, or a peculiar type of spontaneous regression in a tumor that is still actively proliferating.

The occurrence of KS in HIV under ART and with normal or even high CD4 counts has been a matter of concern. Several reports of cutaneous KS in this setting are available, and it would appear that this particular presentation shows a different behavior than the usual form of HIV related KS, with less impact on the global prognosis of the disease [8,9]. Nevertheless, it is important to be aware that this situation may occur also for visceral involvement, and that unusual histologic variants may represent a challenge for pathologist facing small biopsies of unsuspected lesions.

References

[1] Grabar S, Abraham B, Mahamat A, Del Giudice P, Rosenthal E, Costagliola D. Differential impact of combination antiretroviral therapy in preventing Kaposi's sarcoma with and without visceral involvement. J. Clin. Oncol. 2006;24:3408–14. https://doi.org/10.1200/JCO.2005.05.4072.
[2] Lazure T, Plantier F, Alsamad IA, Cabanis P, Malaury E, Blondeau J-R. Bilateral

adrenal Kaposi's sarcoma in an HIV seronegative patient. J. Urol. 2001;166(1):1822–3. https://doi.org/10.1016/S0022-5347(05)65684-4.
[3] Lodi S, Guiguet M, Costagliola D, Fisher M, de Luca A, et al. Kaposi sarcoma incidence and survival among HIV-infected homosexual men after HIV seroconversion. J. Natl. Cancer Inst. 2010;102(11):784–92. https://doi.org/10.1093/jnci/djq134.
[4] Radu O, Pantanowitz L. Kaposi sarcoma. Arch. Pathol. Lab. Med. 2013;137(2):289–94. https://doi.org/10.5858/arpa.2012-0101-RS.
[5] Grayson W, Pantanowitz L. Histological variants of cutaneous Kaposi sarcoma. Diagn. Pathol. 2008;3:31. https://doi.org/10.1186/1746-1596-3-31.
[6] Pantanowitz Dezube BJ, Pinkus GS, Tahan SR. Histological characterization of regression in acquired immunodeficiency syndrome related Kaposi's sarcoma. J. Cutan. Pathol. 2004;31:26–34. https://doi.org/10.1046/j.0303-6987.2004.0132.x.
[7] Burdick AE, Carmichael C, Rady PL, Tyring SK, Badiavas E. Resolution of Kaposi's sarcoma associated with undetectable level of human herpesvirus 8 DNA in a patient with AIDS after protease inhibitor therapy. J. Am. Acad. Dermatol. 1997;37(4):648–9. https://doi.org/10.1016/S0190-9622(97)70188-9.
[8] Chan J, Kravcik S, Angel JB. Development of Kaposi sarcoma despite sustained suppression of HIV plasma viremia. J. Aquir. Immune Defic. Syndr. 1999;22:209–10.
[9] Von Braun A, Braun DL, Kamarachev J, Günthard HF. New onset of Kaposi sarcoma in a human immunodeficiency virus-1-infected homosexual man, despite early antiretroviral treatment, sustained viral suppression, and immune restoration. Open Forum Infect. Dis. 2014;1(1):ofu005. https://doi.org/10.1093/ofid/ofu005.