

## Case Report

# Kaposi sarcoma in an immunocompetent patient: a case report

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### ABSTRACT

Kaposi sarcoma (KS) is an uncommon vascular neoplasm associated with human herpes virus 8 (HHV8) infection. Lesions predominantly present at mucocutaneous sites, but may involve other organs. Epidemiological-clinical forms of KS include classic, endemic, AIDS associated and iatrogenic KS. KS is one of the most common AIDS defining malignancy. Lesions evolve from early (patch stage) macule into plaque stage and nodule stage. Diagnosis is made through clinical, histopathological findings and specific immunohistochemical markers for HHV8. Treatment is either by means of destructive procedures and systemic therapy in case of multiorgan involvement. This report is a case of classic KS in a 74-year nondiabetic, non hypertensive female, with no significant surgical, sexual or history of immunosuppression for the occurrence of KS. This case throws light on addressing KS in an immunocompetent patient or a newer variant of KS yet to be researched upon, which can go clinically misdiagnosed due to lack of significant history.

**Keywords:** KS, HHV8, Vascular neoplasm, Immunocompetent

### INTRODUCTION

Kaposi sarcoma (KS), was first described by Moritz Kaposi in 1872, is a multifocal endothelial proliferation of malignant potential.<sup>1</sup> Though predominantly present at mucocutaneous, can involve other organs also.<sup>10</sup> Four distinct subtypes include; classic, endemic, iatrogenic and AIDS associated.<sup>2</sup> Classic KS starts in lower extremities and progress very slowly. Iatrogenic KS resembles classic KS in presentation, however with a more varied sites of presentation. African endemic form can be locally aggressive but rarely systemically. AIDS associated KS is rapidly progressive and fulminant.<sup>8</sup> HHV8 is considered as causative agent. The entity has gained attention especially after recognition of its association with HIV/AIDS.<sup>10</sup> The strong predisposition among immunocompromised individuals reflects the dependence of the condition upon host immune status. In patients with weak immune system virus can multiply and make susceptible to the condition.<sup>3</sup> Here we report a case of

Classic KS in a 74-year-old healthy female, with no known comorbidities. This case report emphasises on the importance of diagnosing KS in an immunocompetent patient without significant history, using clinical and histopathological knowledge about the condition.

### CASE REPORT

A 74-year-old female, presented with a 6-month history of lesions involving trunk and extremities. Lesions initially started on upper extremities and rapidly progressed to involve trunk and lower extremities in a span of 1 month. Number and size of the lesions progressed with time. Except for the cosmetic appearance the lesions were asymptomatic, with no associated itching or bleeding.

No history of diabetes, hypertension or any other medical history. No recent surgical history, history of blood transfusion or intake of immunosuppressive medications.

No recent sexual exposure history. No recent travel history outside the state. No similar family history.

On physical examination, moderately built and nourished. Vitals were stable. No lymphadenopathy. System examination normal.

Mucocutaneous examination revealed multiple purplish patches, papules and nodules almost symmetrically involving chest, upper extremities and trunk (Figure 1 A and B). Palm and soles spared. Mucosal examination normal. Scalp spared.

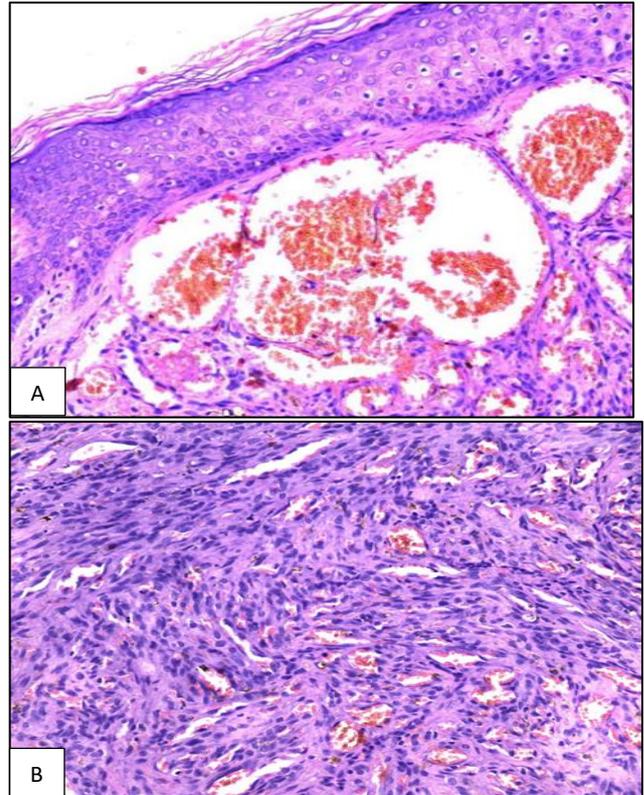
Routine blood investigation and imaging study did not reveal any significant findings.

HIV serology was negative.

Excisional biopsy of the lesion revealed circumscribed tumour arranged in numerous slits like spaces with peripheral ecstatic vessels (Figure 2 A). Spindle cells with moderate nuclear atypia and abnormal mitosis (10-15/hpf) (Figure 2 B). Immunohistochemistry positive for HHV8.

Two sessions of CO<sub>2</sub> laser ablation were done for the lesions (Figure 3).

Patient is under follow up for last 6 months. No recurrence reported till day.



**Figure 2: Histopathology from lesion showing vascular spaces similar to slits with extravasation of RBC and proliferation of spindle shaped cell with cytological atypia.**



**Figure 1 (A and B): Violaceous macules bilateral upper forearms and purplish nodules over mammary region.**



**Figure 3: Post CO<sub>2</sub> laser ablation.**

## DISCUSSION

KS is a rare multifocal tumour with vascular proliferation and four clinical subtypes; classical, endemic, iatrogenic and HIV associated.<sup>4</sup> Classic KS mainly presents in elderly men of Eastern Europe and Mediterranean.<sup>1</sup> Endemic KS is more aggressive and is reported among children and young adults of sub-Saharan Africa.<sup>5</sup>

Iatrogenic variant is more associated with immunosuppressive therapy and transplantation. HIV associated is mostly reported in men having sex with men (MSM).<sup>6</sup> Our case is a rare presentation of KS in an immunocompetent female which can be either a subclassification of classic KS in immunocompetent patients or a newly emerging sporadic variant.<sup>2</sup>

Clinically lesions of KS evolves patch to plaque and to nodule stage.<sup>7</sup> Tumour may ulcerate, cause marked lymphedema or may present as exophytic growths. While KS commonly present in mucocutaneous sites, it can involve lymph nodes as well as visceral organs. Initial patch stage of KS is characterised by small, thin walled, endothelium lined vessels surrounded by large ectatic vessels.<sup>8</sup> The opening of small vascular structure into lumen of more ectatic channels is called 'promontory sign.'<sup>9</sup> Latency associated nuclear antigen HHV8 is most specific immunohistochemical marker, which was positive in our case.<sup>10</sup>

Treatment goals of KS include symptom palliation, prevention of progression and improvement of cosmesis. Limited disease can be treated with ablative procedures and intralesional chemotherapy.<sup>11</sup> In our case CO<sub>2</sub> laser ablative treatment gave promising results.

## CONCLUSION

KS is a low-grade vascular tumour of endothelial origin that is associated with HHV8 infection. The extent and aggressiveness depend on the host immunity. The entity is rarely seen in immunocompetent patients. This case demonstrates the importance of recognising the disease in immunocompetent individual using knowledge of clinical presentation and relating with histopathological and immunohistochemical findings.

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