Unusual presentation of more common disease/injury

Kaposi sarcoma presenting as dermal nodule

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Summary

Kaposi sarcoma (KS) is a rare tumour caused by human herpes virus 8. It is a systemic disease which can present with skin lesions with or without internal involvement. We are presenting here a case of classic subtype of KS in a 60-year-old male who presented with a 4-month history of nodular lesion over right hand.

BACKGROUND

In majority of cases, patients of Kaposi sarcoma (KS) are positive for HIV infections; however, in present case report, the patient was HIV negative and belongs to Sporadic KS that is rare.

CASE PRESENTATION

A 60-year-old male presented with a 4-month history of slow growing nodular lesion on dorsal aspect of right forearm which was 1.5 cm in diameter, purplish in colour and non-tender. Systemic and general examination were unremarkable. Fine needle aspiration cytology revealed sparse scattered spindle cells against haemorrhagic background. A tentative diagnosis of haemangioma was rendered.

Lesion was excised and submitted for histopathological examination, which on microscopic examination showed a dermal nodule (figure 1) with slit like blood-filled spaces lined by spindle cells and filled with red blood cells dissecting the collagen tissue along with occasional mitosis seen (figure 2). Hyaline globules are seen (figure 3A). Appearance of a small vessel protruding into an abnormal vascular space (Promontary sign) and haemosiderin laden macrophages are also seen (figure 3B). Surgical margins of excised specimen were negative for tumour cells. On the basis of above findings, a diagnosis of KS was made. Western blot and ELISA for HIV were negative, and the absolute lymphocyte count was 2.6×10^9 cells/l. The patient remains well at 14 months after initial diagnosis.

Figure 1 Section showing a dermal nodule underlying a normal looking epidermis (H&E×50).

Figure 2 Section showing slit like vascular spaces filled with red blood cells and surrounded by spindle cells (H&E×500).

Figure 3 (A) Section showing hyaline globules in between spindle cells (H&E×500). (B) Section showing promontary sign (black arrow) and haemosiderin laden macrophages (green arrow) (H&E×500).

DIFFERENTIAL DIAGNOSIS

- ▶ Spindle cell haemangioma.
- ▶ Pyogenic granuloma.
- ▶ Angiodermatitis.
- ▶ Bacillary angiomatosis.
- ▶ Angiosarcoma.
- ▶ Kaposiform haemangioendothelioma.

OUTCOME AND FOLLOW-UP

Patient is well till now, 14 months after initial diagnosis without any further intervention.

DISCUSSION

KS was first described by Kaposi in 1872.¹ It is caused by Kaposi sarcoma-associated herpes virus (KSHV), also known as human herpes virus 8 (HHV-8).^{2 3} The virus can be detected in blood of patients before lesions appear.⁴ HHV-8 is one of the few viruses associated with tumourigenesis in humans. This virus causes KS as well as primary effusion lymphoma and some types of multicentric Castleman's disease. The strategies employed by KSHV is by targeting tumour suppressor pathways activated during immune system signalling that may lead to

inadvertent cell proliferation and tumourigenesis in susceptible hosts.

It is a systemic disease which can present with skin lesions with or without internal involvement. Cutaneous lesions vary from macule, patch, plaque, nodule to exophytic lesion.

It is more common in homosexual population, although also seen in heterosexual drug users.⁵ Four subtypes have been described that include classic type, African endemic type,⁶ AIDS related KS and in immunosuppressed patients.⁷

Classic subtype is primarily seen in older males of Mediterranean and eastern European origin. It is commonly seen in patients between 50 and 70 years of age. It has a male predominance. Patients usually present with one or more lesions on extremities, and lesions do not grow rapidly in his subtype.

Fine needle aspirates are usually haemorrhagic or sparsely cellular composed of spindle cells as in our case. Excisional biopsy is needed for making a definitive diagnosis that can delay in treatment.

Histopathological spectrum can be divided roughly in the corresponding clinical variety as macules, plaques, nodules and late aggressive stage. Nodular stage shows a well-defined nodule in dermis. A honey comb like network of blood filled slit like spaces with interweaving spindle cells are present. Endothelial cells lining the vascular spaces are hardly discernible. Mitotic activity is variable but is usually minimal.

Many vascular tumours may cause diagnostic confusion with KS. Our case, however, lacked the large vascular spaces lined by vacuolated cells and lymphoplasmacytic response of a spindle cell haemangioma and the fibrous bands of a Kaposiform haemangioendothelioma. The presence of a spindle cell component excluded bacillary angiomatosis and pyogenic granuloma and the lack of pleomorphism made an angiosarcoma unlikely. Angiodermatitis or pseudo KS can be a close mimic, but it is composed of lobular proliferation of capillaries separated by fibrous tissue instead of spindle cells as in our case.

Incidence of metastasis varies in literature from rare metastasis to less than 10% of KS.⁸ Although about one-third of unselected patients with classic KS reportedly develop another malignancy like non-Hodgkin lymphoma, leukemia or myeloma,^{8–11} this may not apply to HIV-negative KS since a large Italian study found no significant association between HIV-negative KS and lymphoproliferative disorders.¹² Small localised lesions can be surgically excised with a small surrounding margin of normal tissue. Other treatment modalities include low-dose irradiation,¹³ intralesional chemotherapy, interferon α and IgG.^{14 15} For

larger and multifocal lesions, systemic chemotherapy can be used.

Learning points

- ▶ KS should be considered in the differential diagnosis of skin lesions on the extremities, even in HIV negative individuals.
- ▶ Histopathology of the biopsy specimen is the modality of choice for KS. Fine-needle aspirates are unsuitable as first-line investigations for vascular neoplasms.
- ▶ A careful histopathological examination by a pathologist aware of the differential diagnostic features and a high index of suspicion is likely to yield the correct diagnosis in cases without a suggestive clinical background.

Competing interests None.

Patient consent Obtained.

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