a young immunocompromised woman.



A rare case of odynophagia



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Background

Kaposi's sarcoma (KS) is an AIDS defining malignancy. Its aetiology is associated with human herpes virus 8 infection. The disease having been reported to have involved many different bodily sites and despite affecting the head and neck region in 40%-67% of cases, tonsillar KS is extremely rare.1

A rare aetiology of isolated tonsillar mass lesions causing odynophagia was demonstrated in

Case

A young woman who presented to the emergency department complained of having had difficulty swallowing for the past 2 weeks. She had been recently diagnosed with HIV and had not started antiretroviral therapy (ART) in the interim, as her CD4 count was greater than 500. Despite being in good clinical condition, examination of the oral cavity revealed a large membranous lesion occupying the posterior pharynx. On further inspection, the lesion was found to be confined to right tonsillar pillar (Figure 1); multiple cervical lymph nodes were also noted. There were no clinical findings suggestive of cutaneous nor visceral involvement. This was supported by both an unremarkable chest X-ray and an abdominal ultrasound. An excisional biopsy confirmed KS, as evident by the presence of an ulcerated vascular tumour composed of spindle cells, scattered lymphocytes, plasma cells, haemosiderin pigment and erythrocyte extravasation.

Ethical considerations

Permission to conduct the research was obtained from Addington Hospital (Reference number: 9/2/3/R).

Discussion

Despite KS occurring in 1 out of 20 HIV-infected patients in one study and widely known as the second most common tumour related to HIV infection, tonsillar KS is extremely rare.² Although the first case of tonsillar KS was described in the 1970s, there have only been six other case reports in literature thereafter. Three of the cases reported had occurred in confirmed HIVinfected individuals.3,4,5,6,7,8,9 Like the index case, many cases of isolated KS of the tonsils lacked cutaneous involvement further highlighting the importance of performing a biopsy. Although an abscess, lymphangiomatous tumour and a squamous papilloma are some inclusions in the differential diagnosis of a tonsillar mass, the histologic differential diagnoses of Kaposi's sarcoma include angiosarcoma, fibrosarcoma, arteriovenous malformations and spindle cell haemangioendothelioma. Pathologists may often utilise immunohistochemical studies to assist in excluding many differentials.10

The TIS (Tumour, Immune System and Systemic illness) classification for prognosis is still widely utilised, where groups of patients are categorised into 'good' and 'poor' prognosis. Applying the staging to our case would place her in the 'poor' risk category. Although ART remains the cornerstone therapy to the management of KS in HIV infection, other treatment modalities include local palliative radiotherapy, systemic chemotherapy and, recently, the use of targeted agents. Systemic cytotoxic chemotherapy is warranted in patients with advanced or rapidly progressive disease. Newer targeted therapy includes agents that inhibit vascular endothelial growth factor, thalidomide, metalloproteinases and cytokine signalling pathway inhibitors.^{2,11,12} Surgical excision remains a good option for isolated disease. Many of the patients reported to have tonsillar KS had undergone tonsillectomies and were found to be disease free on follow-up years later. Our patient was started

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Source: Photo taken by Yusuf Moolla

FIGURE 1: On examination, a large tonsillar lesion was evident.

on ART, had prompt bilateral tonsillectomy and has performed extremely well on review with no signs of disease recurrence.

HIV prevalence in South Africa may be as high as 12% of the population; although widespread ART roll-out attempts to prevent opportunistic infections, it may change the face of AIDS defining disease and the presentation of malignancies in HIV-infected individuals.¹³ Our case highlights a rare occurrence and emphasises the importance of performing a biopsy when the diagnosis is unclear.

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Competing interests

The authors declare that they have no financial or personal relationships that may have inappropriately influenced them in writing this article.

Authors' contributions

Y.M. was the academic leader for the study and provided the title, abstract, discussion and overall editing of the article. L.N. contributed to the case study by providing the description and assisting in histology findings.

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