Kaposi’s Sarcoma of an Intra-Parotid Lymph Node in a HIV-Negative Patient

SUMMARY

Background. Kaposi's sarcoma (KS) as one of the defining tumours of AIDS, was described as multiple slowly progressing pigmented skin plagues and as vaso-formative lesion in microscopic finding. Several forms of the disease have been suggested, such as mucocutaneous and lymph nodal. KS is rarely seen in the major salivary glands. Furthermore, KS of parotid tissue or intra-parotid lymph node is extremely rare in HIV-negative patients.

Case Report. We report a case of a right parotid mass as an early sign of KS infection in a 57-year-old patient. The problems related to the diagnosis, the management strategy of such a rare condition and prognosis are also discussed. Complete surgical excision is suggested, followed by adjuvant radiotherapy and management of any other suspicious lesions confirmed by clinical and histo-pathological examination.

Conclusions. KS is a rare tumour of the parotid gland but practitioners need to be reminded of rare cases in their differential diagnosis.

Keywords: Kaposi's Sarcoma; Intra-Parotid Lymph Node; HIV Infection

Introduction

Kaposi’s sarcoma (KS), a cutaneous malignancy of lymphatic endothelial cells, was originally described by Moritz Kaposi in 1872. Since his original description, 4 new forms of the disease have been suggested:

- sporadic;
- transplantation associated;
- endemic African;
- epidemic, acquired immunodeficiency syndrome (AIDS)-related.

The sporadic or classical KS lesions usually are slowly progressive, involving the skin around the angles, the legs, the hands and arms to a lesser extend and frequently the lymph nodes draining those areas. The course of the disease is generally indolent, and the patients survive an average of 10-15 years. The transplantation associated or iatrogenic KS form is found among allograft recipients with fatal course, but spontaneous regression may be observed if immunosuppression is removed. The endemic form is occurred in African adult males and children. Extra-cutaneous involvement in the endemic form is usually associated with an extremely poor prognosis. The epidemic KS is found among patients with acquired immunodeficiency syndrome (AIDS) and has experienced a remarkably increased prevalence. KS is one of the defining tumours of AIDS, and is rarely seen in the major salivary glands. However, KS of parotid tissue or intra-parotid lymph node is extremely rare in a non-immunocompromised and HIV-negative patient.

We present a case of a right parotid mass as an early sign of KS, in an HIV-negative patient. The problems related to the diagnosis, the management strategy of such a rare condition, and prognosis are also discussed.

Case Report

A 57-year-old Caucasian male was referred for evaluation of a painless mass on the right parotid gland. The mass was firm, non tender and smooth on palpation. No palpable cervical lymph nodes were found, and the evaluation of parotid gland function didn’t indicate any diminishing of salivary flow. The mass was painless for almost 2 years, and only recently (the last 6 months) increasing in size.
Taking into consideration the radiological and clinical evaluation, an adequate parotidectomy was performed. The mass was excised completely and the remaining parotid tissue was clinically normal (Fig. 3).

The histological examination of the lesion was indicative of KS. The macroscopic findings revealed that the tumour was a well circumscribed grey-brown mass and was located within the substance of the parotid gland. The histology of the tumour was similar to the plaque-stage KS of cutaneous lesions. It was well defined, surrounded by fibrous tissue varying in thickness, with anastomotic branches of spindle shaped tumour cells in vaso-formative pattern. Lymphocytes and plasmacytes were found within the spindle cells, whereas numbers of lymphocytes and 1-2 lymph nodules were identified in the periphery, resembling a lymph node occupied by the tissue described above. Extravasated erythrocytes were abundant within the slit-like vascular spaces. Characteristic eosinophilic globules, PAS positive, were found occasionally (Figs. 4 and 5). The immunohistochemical analysis revealed spindle cells reacted with factor VIII-related antigen, CD 31, CD 34 and vimentin, confirming the vascular origin of the tumour. The microscopic findings and immunohistochemical results confirmed the diagnosis of KS.

In addition, HIV antibodies were negative in the follow-up evaluation, whereas serum positivity for HHV-8 antibodies was tested by the ELISA method.

Figure 1. Sonography of right parotid gland with lesion

Figure 2. MRI scan that shows a solid lesion on the right parotid gland (arrow)

Figure 3. The excision of the intra-parotid tumour

Figure 4. Branches of spindle shaped cells in vaso-formative pattern (H.E. x 400)
The treatment of KS is a stage and clinical form depended procedure. Surgical resection and adjuvant radiotherapy may be proved adequate in most of the cases.

Adjuvant radiotherapy of 2000 cGy was administered 1 month postoperatively, and one year after treatment the patient was alive and in good general health, with no evidence of the disease, and serologically negative to HIV antibodies.

Discussion

KS is considered to be a virus-associated multifocal neoplasm. It develops with multiple reddish purple maculae in the skin, many of which evolve into plaques and finally subcutaneous nodules.

There are a number of AIDS-defining diseases including malignancies, of which KS is one of the more specific. Therefore, KS is likely to be included in the differential diagnosis of a variety of head and neck, and more specifically, salivary gland presentations of HIV infected patients. Although lymph node involvement may occur in all 4 clinical forms, and sometimes can precede the development of skin lesions or may even occur in their absence, it is more frequently seen in the AIDS-related form.

KS-associated herpes virus (KSHV) is believed to play an etiologic role in the development of KS in patients, either with or without evidence of HIV infection. In 1994 Chang et al discovered a previously unknown KS-associated herpes virus, the human herpes virus type 8 (HHV-8), in virtually every KS lesion examined. KSHV now is believed to be the primary cause of all types of KS. HHV-8 also is believed to be transmitted sexually and to precede the development of KS. Additional studies have shown that antibodies to HHV-8 are present in approximately 90% of patients with KS.

In the present case, there was no evidence of HIV positive antigens, postoperatively and in the follow-up, but HHV-8 antigens were positive of infection, while histopathologic and immunohistochemical examination confirmed the diagnosis of KS. A thorough dermatological examination showed no other evidence of the disease. In addition, chest and upper and lower abdomen MRI revealed no other evidence of KS, so the parotid lesion in our patient could be considered as a primary KS of the parotid gland (Figs. 6 and 7).

The treatment of KS is a stage and clinical form depended procedure. Surgical resection and adjuvant radiotherapy may be proved adequate in most of the
cases. There have been reports for advanced disease or un-resectable lesions to be treated with radiation therapy alone or with concomitant chemotherapy which includes vincristine, bleomycin, etoposide or vinblastine. Our patient received adjuvant radiotherapy of 2000 cGy, 1 month postoperatively. In the 6 and 12 month follow-up, the patient has been free of the disease.

KS is rarely related with poor prognosis. Instead, patients with KS in the epidemic AIDS form, usually succumb to infectious compilations of AIDS. Therefore, most HIV-positive patients affected by KS have a poor prognosis, and an infectious disease specialist is decisive for the initiation of specific therapy. This includes highly active antiretroviral therapy (HAART) and prophylactic antibiotic administration.

In conclusion, although KS is a rare tumour of the parotid gland, especially in HIV-negative patients, practitioners need to be reminded of such cases in their differential diagnosis.

References


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