UNUSUAL PRESENTATION OF OROPHARYNGEAL KAPOSI’S SARCOMA

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Abstract: Background. The aim of this article is to focus ear, nose, and throat (ENT) practitioners on a pathology rarely seen within the head and neck mucosa.

Methods. A 62-year-old black African man was referred to our ENT department for dysphagia and hoarseness. Physical ENT examination revealed a smooth normal mucosal mass on the left lateral pharyngeal wall, which masked the inlet of the larynx, and bilateral cervical lymph nodes but no other mucosal lesions. Anti–human immunodeficiency virus antibodies were found to be negative. Hemoglobin, hematocrit, white blood cell count, and platelet count were normal. Serum protein electrophoresis was also normal. A CT scan confirmed the lesion and lymph node involvement. Treatment consisted of a transoral endoscopic resection with ligature of the pedicle, with the patient under general anesthesia. The patient’s improved medical condition permitted 11 cycles of bleomycin (15 mg/3 weeks).

Results. A 5-month fibroscopic follow-up control showed no recurrence, and total clinical regression was observed after 1 year. Three years later, all ENT lesions had disappeared, with no adenopathy or mucosal lesions.

Conclusion. Kaposi’s sarcoma is a pathology that the ENT practitioner must keep in mind when patients present with a laryngopharyngeal mass. The symptomatic cases were all surgically treated; however, death subsequently occurred.

Keywords: endemic; Kaposi’s sarcoma; oropharynx

Kaposi’s sarcoma (KS) is now a well-defined pathology. Its most frequent form is cutaneous or mucosal associated with acquired immunodeficiency syndrome (AIDS). Although lymph node involvement may occur in some cases, it can precede the development of skin lesions. Oropharyngeal lesions are most frequently encountered on the tonsils or the base of tongue and rarely distant from the lymphoid structure. We report an unusual case of an airway obstruction because of a pediculed mass in a patient with endemic non-AIDS KS.

CASE REPORT

A 62-year-old black African man was referred to our ear, nose, and throat (ENT) department for dysphagia and hoarseness. The patient had been in France since he was 25 years old.
The patient also had a previous medical history of high blood pressure, hepatitis C, and a cerebral ischemia with right hemiplegia sequelae and aphasia 2 years before admission. This included a cervical lymph node biopsy performed in our ENT department 2 years prior to admission, with a KS diagnosed on histologic examination. No previous history of smoking, alcohol abuse, homosexuality, blood transfusion, or intravenous drug abuse was observed. The KS was diagnosed in 2001 on the basis of biopsy of skin and mucosal lesions.

Physical ENT examination revealed a smooth normal mucosal mass on the left lateral pharyngeal wall, which masked the inlet of the larynx (Figure 1), and bilateral cervical lymph nodes but no other mucosal lesions. This mass did not present the classic hemangioma-like aspect of KS lesions.

Anti–human immunodeficiency virus antibodies were found to be negative on 3 occasions using the enzyme immunoassay method. Hemoglobin (8.4 mmol/L), hematocrit (0.41), white blood cell count, and platelet count were normal (273 Giga/L). Serum protein electrophoresis was also normal.

A CT scan confirmed the lesion and lymph node involvement. The oropharyngeal lesion was on a pedicle on the lateral left pharyngeal wall with no contact at the base of tongue or the tonsils. The enhancement was major and homogeneous (Figure 2). Multiple cervical adenopathies with homogeneous enhancement were noted.

Treatment consisted of a transoral endoscopic resection with ligature of the pedicle under general anesthesia (Figures 3 and 4).

The histologic analysis of the surgical specimen revealed a tissue mass with no lymphoid tissue. Epithelial tissue covered the entire mass. The specimen presented a proliferation of spindle cells with large monomorphic hyperchromatic nuclei and numerous cosinophilic inclusions. A vascular component containing red blood cells was observed. Immunohistochemical examination of spindle cell proliferation showed positivity for
cluster of differentiation molecule 31 (CD31) and human herpesvirus 8 (HHV8) (Figure 5). The pedicle and the margin were tumor free.

A 5-month fibroscopic follow-up control showed no recurrence. However, during this period, right tonsil and left vallecula lesions appeared. At initial diagnosis, no general treatment was provided because the patient had a poor medical status. Following the diagnosis of the right tonsil and vallecula lesions, the patient’s improved medical condition permitted the use of chemotherapy. Eleven cycles of bleomycin (15 mg/3 weeks) was delivered. Complete clinical regression was observed after 1 year. Three years later, all ENT lesions had disappeared, with no adenopathy or mucosal lesions.

DISCUSSION

KS was first described in 1872 by the Hungarian dermatologist, Moritz Kaposi. The etiology of KS remains the subject of further investigation (ie, the reactive hyperplasia triggered by angiogenic factors as well as the causative role of the HHV 8). HHV8 could be present in all KS lesions, and cumulating data strongly suggest that HHV8 is involved in the pathogenesis of KS.

KS is currently classified into 4 types: classic, which primarily affects elderly men of Mediterranean and Eastern European origin; epidemic or AIDS associated; acquired or transplant-related; and endemic, which is common in parts of Africa. A more aggressive pathology is often seen in infectious (HIV) immunosuppressed patients. KS particularly involves the skin, the lymph nodes, and organs such as the gastrointestinal tract. In ENT pathology, the neoplasm involves the mouth, the lymph nodes, and the oropharynx. Buccopharyngeal lesions are found in 10% to 50% of cases. The preferential localizations in the mouth and in the oropharynx are the palate and the Waldeyer’s ring, respectively. These mucosal lesions are usually multiple reddish-purple macules or papules.

KS in non-AIDS patients is associated with an indolent course. In these non-AIDS forms, oral involvement is rare, and fewer than 15 cases have been reported in the literature. Endemic KS is found in all age categories and occurs more often in men. This endemic non-AIDS form of KS has been confined to Northern Europe to Africa. Pathologic diagnosis is difficult.
because of similarities between KS and vascular tumors. The localization, macroscopic characteristics, pattern progression, and histologic findings of the lesions are important for KS diagnosis. However, there is a substantial argument for the detection of HHV8.

For Emery et al8 on barium pharyngography, well-defined nodular lesions with no mucosal ulceration can be specific, and CT scan may be useful to assess deep soft tissue involvement.9

Standard treatment consists of systemic chemotherapy or immunotherapy. Several chemotherapy agents (vinblastine, vincristine, etoposide, interferon-α) have been recommended. This systemic treatment has been reserved for cases of diseases with multivisceral involvement.11

Radiotherapy has been widely used in doses ranging 600 to 3000 cGy. This approach has halted the growth and has produced complete regression in up to 70% of cases.12 For Jindal et al the “standard-dose” for KS in patients with AIDS (1500 cGy) has been ineffective and can be increased to 4800 cGy. A period of watchful waiting can be proposed to asymptomatic ENT localization.9 Some authors, owing to positive herpes serology, propose treatment with protease inhibitors.13 Benfield et al13 presented 3 cases of patients with complete remission following treatment with protease inhibitors with a 3- to 4-month posttreatment follow-up. Local therapy can also be an effective approach in management of symptomatic KS lesions: direct surgical excision, laser therapy, photodynamic therapy, cryotherapy, and local chemotherapy.11,14–16

This case report not only presents a rare clinical occurrence of KS but also demonstrates the possibility of aggressive KS in a non-AIDS patient. Only 2 KS cases with airway obstruction have been reported in the literature.17,18 These 2 patients died shortly after a tracheotomy secondary to local hemorrhage. The transoral approach was preferred to avoid the hemorrhagic risk (surgical ligation of the pedicle) (Figure 5). With regard to cases of symptomatic airway lesions, surgery affords immediate benefit to the patients compared with the slower medical treatment approach. In contrast, the treatment of the other lesions requires a case-by-case assessment.

CONCLUSION

KS is a pathology that the ENT practitioner must keep in mind when patients are seen with a laryngopharyngeal mass.

Our case of KS in a non-AIDS patient was unusual because of the delay between lymph node and oropharyngeal localizations and the lesion’s position far from Waldeyer’s ring. Pathologic diagnosis remains difficult; however, there is increasing evidence that HHV8 detection could be useful in achieving a final diagnosis. We performed a primary excision owing to the unusual characteristics and symptoms. In patients with AIDS, life expectancy remains limited. This single surgical treatment approach of the symptomatic lesions permits an improvement in the quality of life with relatively low risk.

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REFERENCES


