given a physical examination and a full medical history should be taken. Hyperandrogenism and insulin resistance should be ruled out whenever these appear to be present. As far as we are aware, no association with hypochondroplasia has been previously reported. The characteristic phenotype of this disease means that a general physical examination will be sufficient to guide the choice of complementary examinations given to patients.

References


Primary Kaposi Sarcoma of the Penis in an HIV-Negative Patient

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To the Editor:

Kaposi sarcoma is a vascular tumor of multifocal origin first described by Moritz Kaposi in 1872. In the Mediterranean area, the classic form is mainly seen on the lower limbs of the elderly. Isolated involvement of the penis is rare, and although it is seen in AIDS patients—where it is the initial manifestation in 2-3% of cases—it is extremely uncommon in human immunodeficiency virus (HIV)-negative patients. In the last 20 years, only 15 cases of immunocompetent patients with primary Kaposi sarcoma of the penis have been described in English-language journals.

We present the case of an 80-year-old man, with no relevant history, presenting a rapidly growing asymptomatic tumor on the penis that had developed 2 weeks earlier. The patient had no history of local trauma, immunosuppression, intravenous drug addiction, blood transfusions, or homosexual acts. Physical examination revealed a soft pink pedunculated nodule of 10 mm in diameter on the coronal sulcus. He also presented a second clearly circumscribed painless red-violaceous lesion of 4 mm in diameter that had been present for several years (Figure 1). There was no evidence of inguinal gland involvement, hepatosplenomegaly, or other
mucocutaneous lesions. Histological studies of the larger lesion showed a multinodular tumoral proliferation made up of fusiform cells with lengthened hyperchromatic nuclei and occasional mitosis. Bundles of these cells formed disruptions containing red blood cells (Figures 2 and 3). Several areas of eosinophilic globular bodies and hemosiderin deposits could also be seen. Immunohistochemical staining proved positive for CD31 and CD34. On removal, the second lesion showed similar histopathological abnormalities. Complete blood count, coagulation, and T-cell count results did not provide any relevant findings, and the patient was seronegative for HIV in 2 tests. Primary Kaposi sarcoma of the penis in an immunocompetent patient was the final diagnosis. A year later, the patient remains stable with no further lesions.

The pathogenesis of Kaposi sarcoma is unknown, although the epidemiological characteristics probably indicate an infectious cause. Human herpes virus type 8 (HHV8) is implicated in vascular hyperplasia, but while a necessary factor, it is not sufficient cause in itself. The high seroprevalence of HHV8 in individuals with high risk sexual activity would appear to support this form of transmission in adults; however, the detection of HHV8 antibodies in children suggests there are other nonsexual means of transmission, probably through saliva.1,2

Kaposi sarcoma of the penis is clinically identified by painless reddish-violaceous colored nodules. Other less common forms of presentation include multiple papules, plaques, or rapidly growing pedunculate tumors.1 Most cases consist of 1 or 2 isolated lesions1 most commonly found on the glans, although the foreskin, coronal sulcus, urethral meatus, and scrotum can also be affected.1

The histological pattern for Kaposi sarcoma of the penis is similar to that seen in other anatomical locations. In the tumor phase, nodules containing a network of blood-filled vascular spaces appear along with fascicles of fusiform cells. These fusiform cells have a clearly defined cytoplasm and an ovoid nucleus and are characteristically CD34 positive.6 In some areas, differential diagnosis with pyogenic granuloma and spindle-cell hemangioendothelioma might be necessary. Spindle-cell tumor proliferation with the presence of abnormal cells and the formation of disruptions would not support a diagnosis of pyogenic granuloma. Spindle-cell hemangioma would be clearly shown by marked cytoplasmic vacuolization and an absence of atypical cells.7

Primary Kaposi sarcoma of the penis can be treated by local surgery, radiotherapy, electrocoagulation, laser therapy, and injection of interferon α into the lesion, although there are no established treatment guidelines. Surgery is recommended for small or solitary lesions. Radiotherapy is used in larger lesions and systemic chemotherapy is reserved for more advanced cases with visceral involvement or widespread lesions. The clinical course of primary Kaposi sarcoma of the penis is variable and local recurrence is uncommon if the primary tumor is completely eliminated.3

We would like to stress that although primary KS of the penis is extremely uncommon in immunocompetent subjects it must be considered in the differential diagnosis of nonspecific lesions in the genital area. Histological examination is advisable in unclear cases where no clinical characteristics are available to establish a definitive diagnosis.

References

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Figure 2. Tumor composed of a network of vascular spaces with a proliferation of fusiform cells (hematoxylin-eosin × 20).

Figure 3. Detail of the vascular proliferation showing the presence of hyaline globules (hematoxylin-eosin × 400).