Eosinophilic Granuloma of the Vulva

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A 46-year-old woman sought treatment of vaginal pruritus of 6 months' duration. She also was bothered by generalized skeletal aching that was most prominent in her legs. The patient had a history of hypertension.

Laboratory studies, chest films, and bone scan findings were within normal limits. A 0.5- x 0.5-cm nodule was found on the vulva. Histologic examination of a biopsy specimen from the lesion revealed Langerhans' cells that suggested eosinophilic granuloma. This diagnosis was confirmed by electron microscope examination of the specimen that demonstrated abundant Birbeck granules (A, arrows) in the cells. B-cell, T-cell, and macrophage markers were absent; the specimen was positive for S-100, CD1a, and leu 22 (CD43) markers. These findings are consistent with Langerhans' cells.

A CT scan indicated a possible residual vulval mass. Subsequent deeper excision and examination of the vulval nodule did not yield additional pathologic conditions.

Drs Hesham Taha, K. Economos, S. Fiasconaro, J. C. Vuletin, G. Kostandy, and David Dosik of New York Methodist Hospital, Brooklyn, write that eosinophilic granuloma is a non-neoplastic lesion of unknown origin. It is characterized by an intensive proliferation of the reticulohistiocytic element with varying numbers of eosinophils, leukocytes, neutrophilic leukocytes, lymphocytes, plasma cells, and multinucleated cells. The lesion can occur in unifocal or multifocal form. Eosinophilic granulomas generally affect bone; involvement of the female genital tract is very rare. When the lesion does occur in the genital tract, it is found almost exclusively in the squamous epithelium. Patients are typically younger than 20 years; men are approximately twice as likely as women to have the disease. A study of 238 cases of eosinophilic granuloma found that 35% of patients were women. Unifocal lesions were noted in 65% of all cases, with the skull, femur, ribs, jaw, and other bones the predominant sites. Nine percent of patients presented with organ or soft tissue involvement (lung, lymph nodes, skin, and submaxillary glands). Only two women demonstrated vulval lesions; these patients had multifocal disease with additional nodules on areas including the skin, mastoid bone, mandible, and buccal mucosa. The doctors state that their research yielded only one other report of eosinophilic granuloma of the vulva.

The course of individual lesions is extremely variable, and local recurrences may occur. Management of unifocal disease includes radiographic survey of the involved area and a bone scan to determine the extent of the disease followed by surgical resection of the isolated lesion. Supportive therapy for pain and observation for the development of other lesions usually is needed. Radiographic evaluation of the site of the original involvement should be done after initial excision and then every 6 months for 1 to 2 years.

REFERENCES:


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