uli. It is unclear whether this excretion causes luminal blockade or clears the cytoplasm of the inclusions. Furthermore, some eccrine glands undergoing degeneration showed complete replacement of their normal cytoplasmic constituents by the inclusions. We believe that these morphologic findings are causally associated with anhidrosis and that anhidrosis, even without the simultaneous presence of the characteristic angiokeratoma corporis diffusum or corneal opacities, should be considered as a possible sign of Fabry's disease.

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Malignant papillary mesothelioma of the tunica vaginalis testes: Cutaneous metastases showing pagetoid epidermal invasion

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Malignant papillary mesothelioma of the tunica vaginalis testis is an extremely rare tumor arising from the mesothelium of the tunica vaginalis. A total of 14 cases have been reported, with only two exhibiting cutaneous metastases. We present a third case of cutaneous metastases arising from this unusual tumor. In addition, this is the first reported case of the occurrence of pagetoid epidermal invasion in this condition. (J AM ACAD DERMATOL 1987;17:887-90.)

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Mesotheliomas are tumors arising from mesothelial cells lining the serous membranes of the pleura, pericardium, and peritoneum. During fetal development the tunica vaginalis propria testis is

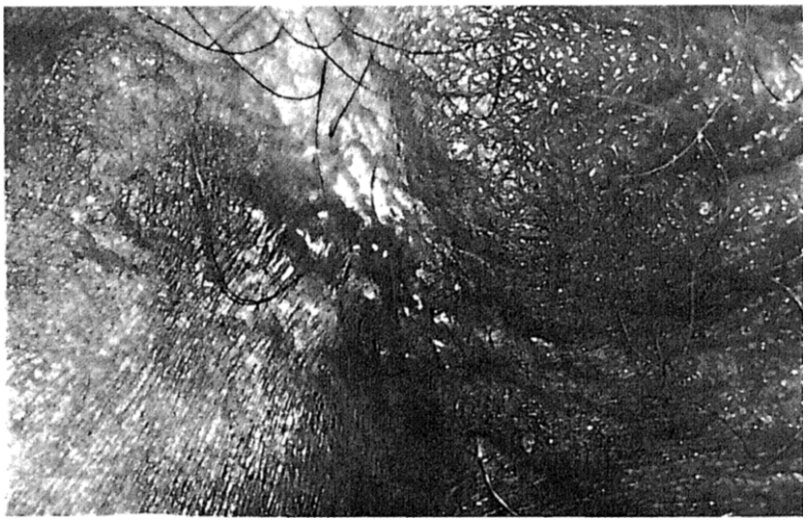

Fig. 1. Firm, waxy, papular cutaneous metastases at the junction of the proximal penis and the scrotum.

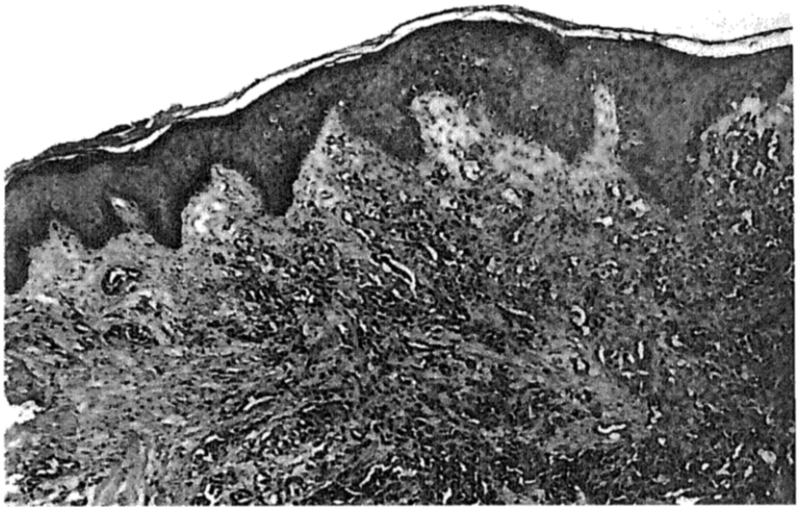

Fig. 2. Photomicrograph of biopsy specimen showing cutaneous metastasis with multiple clefts and tubuloglandular structures in the papillary and reticular dermis. (Hematoxylin-eosin stain; ×25.)

formed from an outpouching of the abdominal peritoneum and thus has a mesothelial lining. Mesotheliomas may therefore rarely arise in the tunica vaginalis propria testis.[1] They may occur as papillary or cystic tumors and may be localized or diffuse in their pattern of growth. Localized mesotheliomas are often benign; their diffuse counterparts are usually malignant.[2,3]

To date there have been 14 reported cases of malignant papillary mesothelioma of the tunica vaginalis propria testis, but cutaneous involvement has been noted only twice.[2,3] We report an additional case of this rare tumor and emphasize the histologic finding of pagetoid epidermal invasion, which has not been previously noted.

CASE REPORT

A 49-year-old white man had undergone a Lord procedure for a right hydrocele in July 1982. Scrotal exploration and biopsies of the testis and tunica vaginalis

showed no evidence of tumor. In November 1983 a firm, nontender mass of the upper pole of the patient's right testis was noted. Several courses of systemic antibiotic therapy for a presumed epididymitis were without effect. In September 1984 the patient underwent a right orchiectomy and hemiscrotectomy, at which time the diagnosis of malignant papillary mesothelioma of the tunica vaginalis propria testis was made. Right inguinal node dissection revealed nodal metastasis in 5 of 10 inguinal nodes sampled. A right radical retroperitoneal node dissection was negative. Chest radiographs and computed tomography (CT) scans revealed multiple parenchymal lung nodules, which on CT guided-needle biopsy proved to be metastatic mesotheliomas.

The patient was admitted for chemotherapy in January 1985, and several 1-3 mm nontender, firm, waxy papules were noted on the penis and scrotum (Fig. 1), as well as a tender 1 × 2-cm indurated plaque on the ventral aspect of the penis. Histopathologic and electron microscopic examination of tissue from the papules and plaque showed findings consistent with malignant papillary mesothelioma. Treatment with multiple courses of chemotherapy have been unsuccessful at halting further tumor growth.

Histopathologic study. Pathologic examination of skin biopsy specimens from the penis and scrotum showed clefts lined by a single or double layer of variably sized cuboidal cells forming tubuloglandular structures throughout the papillary and reticular dermis (Figs. 2 and 3). In some areas these cells lay over thin, branching connective tissue stroma and formed delicate papillary structures. The cuboidal cells stained positively with periodic acid–Schiff reagent, were resistant to diastase digestion, and did not stain with mucicarmine. Stains with immunoperoxidase-labeled antikeratin antibodies were positive, but they were negative for carcinoembryonic antigen, β-human chorionic gonadotropin, and α-fetoprotein. These findings are consistent with the diagnosis of malignant papillary mesothelioma.[4-6] An unexpected finding was the presence in the suprabasal layer of the epidermis of numerous large cells with abundant pale-staining cytoplasm, prominent nucleoli, and mitoses. These atypical cells extended upward into the malpighian layer in a pagetoid fashion (Fig. 4).

Electron microscopy. Transmission electron microscopy of the skin biopsy specimens demonstrated aggregates of two cell types. The first type consisted of pale-appearing cells with large smooth bordered oval nuclei containing finely coarse chromatin and prominent nucleoli. They had abundant cytoplasm, few mitochondria, short profiles of rough endoplasmic reticulum, and occasional tonofilaments near the nuclear border. The second cell type appeared dark with irregularly shaped, highly indented nuclei containing coarse chromatin and prominent nucleoli. The cytoplasm contained numerous mitochondria, prominent Golgi apparatus, abundant irregularly dilated rough endoplasmic reticulum, and thick bundles of tonofilaments closely apposed to the nuclear border. Elongated microvilli extended from the surface of both cell types, and desmosomes and tight junctions were present between cells. No intracellular laminae were seen, but a thin basal lamina, sometimes interrupted, surrounded groups of cells. These findings are also consistent with a diagnosis of malignant papillary mesothelioma.[4,7,8]

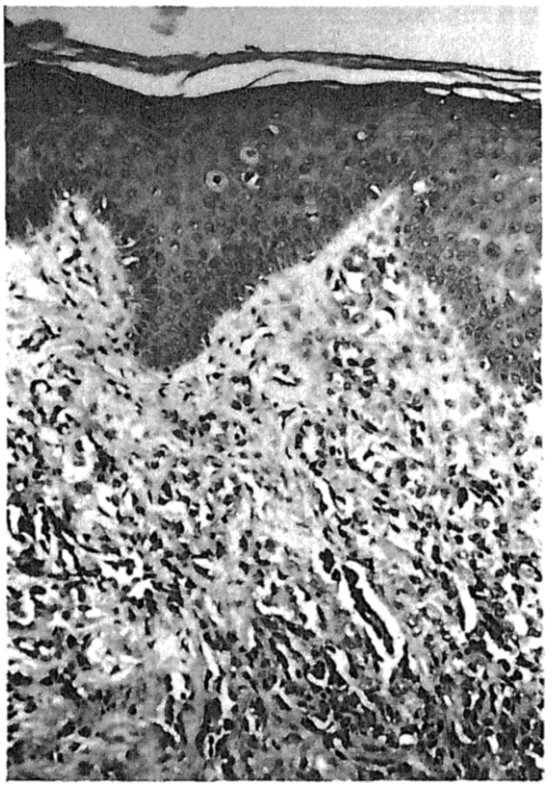

Fig. 3. High-power view showing clefts and glandular structures lined by variably sized cuboidal cells. (Hematoxylin-eosin stain; × 40.)

DISCUSSION

Clinically, malignant papillary mesothelioma of the tunica vaginalis propria testis presents as an asymptomatic, firm, intrascrotal mass associated with a hydrocele. Solitary, clinically benign variants of mesothelioma, also known as adenomatoid

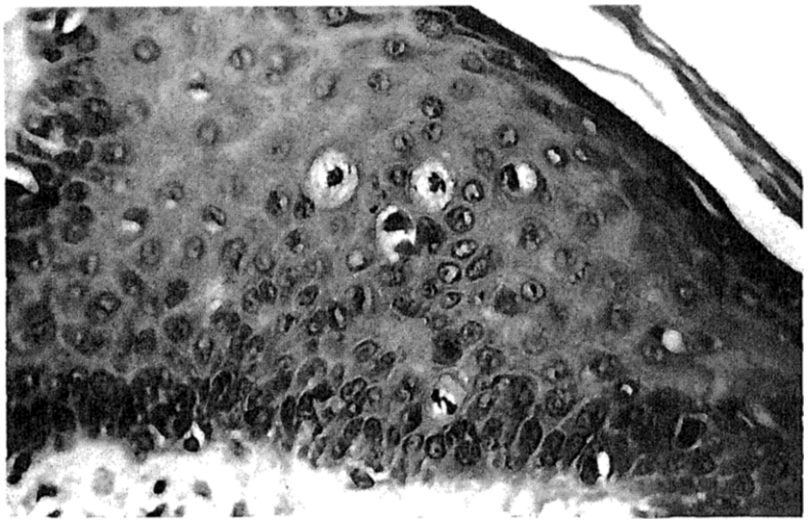

Fig. 4. Photomicrograph of epidermis specimen showing a pagetoid distribution of large, pale-staining cells. A mitotic figure is present. (Hematoxylin-eosin stain; × 100.)

tumor, may also originate in the tunica vaginalis propria testis.[4,7,8] Treatment for both forms is surgical excision. To date only 14 cases of malignant papillary mesothelioma of the tunica vaginalis propria testis have been reported, and only twice has cutaneous involvement been noted.[2,3] The case reported here is the third with invasion of the skin.

Diagnosis was made on histopathologic, histochemical, and electron microscopic grounds and met the diagnostic criteria proposed by Japko et al.[1] This case is most remarkable for the presence of pagetoid epidermal invasion on histopathologic examination, a finding not previously reported. Other tumors exhibiting pagetoid growth are superficial spreading melanoma in situ, superficial spreading malignant melanoma, Bowen's disease, bowenoid papulosis, Paget's disease of the breast, extramammary Paget's disease secondary to adenexal carcinoma, gastrointestinal tract adenocarcinoma, and urinary tract endothelial carcinoma.[9,10] Thus malignant papillary mesothelioma of the tunica vaginalis propria testis must also be considered in the differential diagnosis of tumors causing pagetoid epidermal change.

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