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Giant Tumor of the Back

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REPORT OF A CASE

A 47-year-old white man presented with a 6-month history of what he thought was a cyst on his left upper back area (**Figure 1** and **Figure 2**). The cyst had been foul

smelling for some time and had recently begun to bleed. His medical history was significant for schizophrenia. His only medication was chlorpromazine.

Physical examination revealed an 8.0 × 9.0 × 4.5-cm multiloculated, ulcerated, necrotic, foul-smelling

tumor attached by a narrow pedicle to the left upper scapular area. An excisional biopsy was performed (**Figure 3** and **Figure 4**).

What is your diagnosis?



Figure 1.

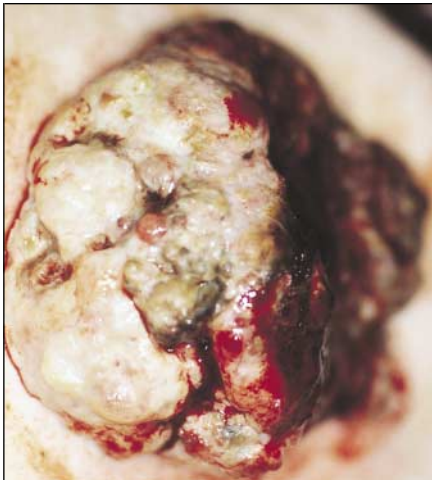


Figure 2.

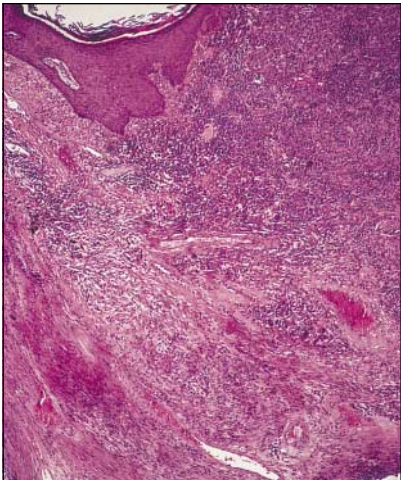


Figure 3.

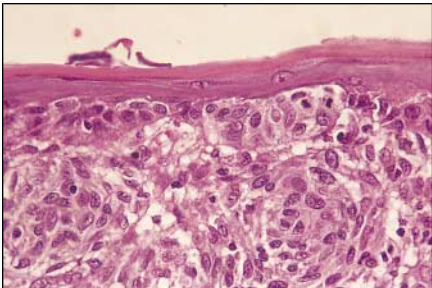


Figure 4.

Recurrent Nodules on the Feet of a Child

Adam Rotunda, MD; Deborah Schappell, MD; Leslie Robinson-Bostom, MD; University of California, Los Angeles (Dr Rotunda), and Brown University School of Medicine, Providence, RI (Drs Schappell and Robinson-Bostom)

REPORT OF A CASE

A healthy 8-year-old girl presented with a 4-year history of several nodules on her feet. She had no significant medical problems other than a history of eczema as an infant. She was not taking any medications. There was no family history of similar lesions. The initial lesion was excised 4 years earlier, then recurred, and has since

resolved. A general pathologist initially interpreted the pathologic findings as a hypertrophic scar. There has been an increase in the number of lesions since the first one was noticed, but the lesions have since remained unchanged in appearance. The patient reports that pressure from her shoes causes her pain.

Physical examination revealed several discrete, firm, pink, dome-shaped nodules ranging from 6 to 9 mm in

diameter distributed on the lateral aspect of the left foot, medial aspect of the left fifth toe, and dorsal aspect of the right fifth toe (**Figure 1**). A hyperpigmented scar was noted on the lateral aspect of the left foot. A biopsy specimen from the initial lesion was reviewed (**Figure 2** and **Figure 3**).

What is your diagnosis?



Figure 1.

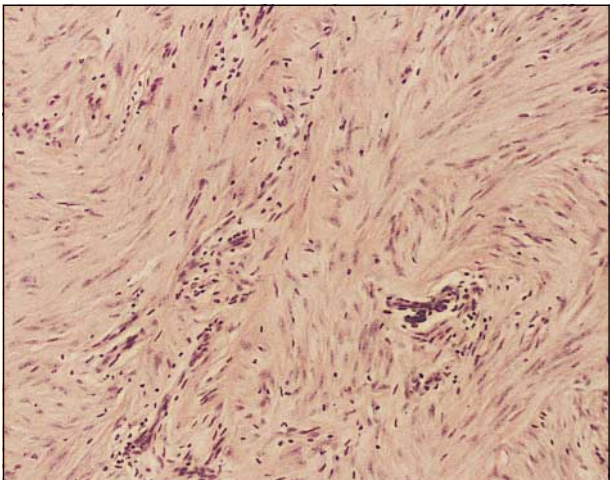


Figure 2.

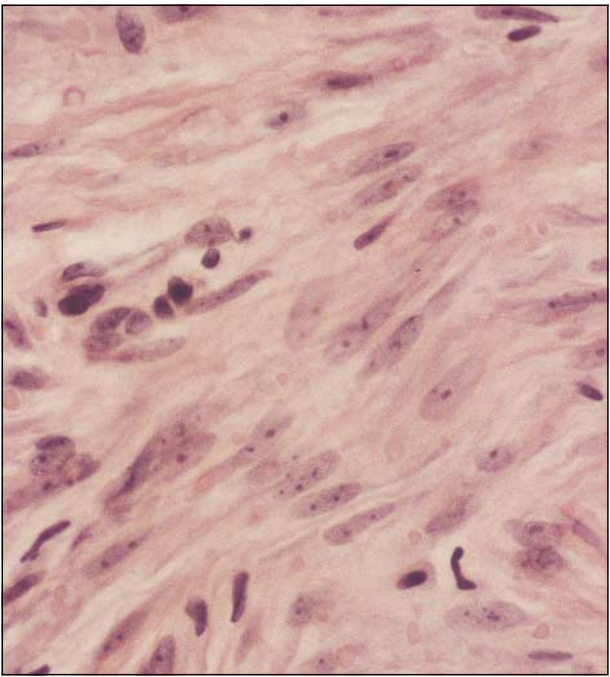


Figure 3.

Vulvar Lesion in a 45-Year-Old Woman

Sophie Frayssé-Consigny, MD; Olivier Chosidow, MD; Pierre-André Becherel, MD; Annick Datry, MD; Camille Frances, MD; Hôpital Pitié-Salpêtrière, Paris, France

REPORT OF A CASE

A 45-year-old white woman presented with a 2-year history of a persistent warty lesion of the vulva accompanied by dysuria and recurrent infection of the urinary tract. She had been unsuccessfully treated with topical acyclovir.

Physical examination disclosed a painless, papillomatous nodule on the left labium minus and some infiltrative perianal papules and nodules (**Figure 1**). The patient had left inguinal lymphadenopathy. Her abdomen was tender to palpation. She was otherwise well. There was no hematuria. The results of the following laboratory parameters were within normal limits: complete blood cell count, serum electrolyte profile, serum calcium levels, liver function tests, urinalysis, and screening for immunodeficiency virus and syphilis. A biopsy specimen was obtained from the vulva (**Figure 2**).

What is your diagnosis?



Figure 1.

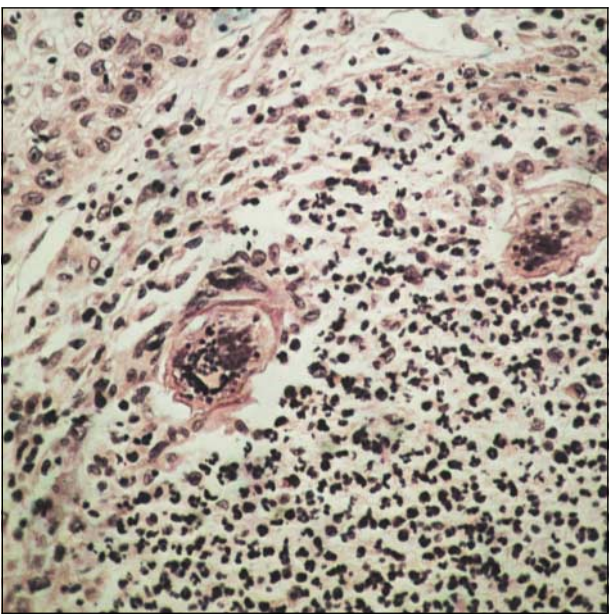


Figure 2.

Erythematous Rash on the Chest

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REPORT OF A CASE

A previously healthy 28-year-old white woman presented with a 6-month history of a nonpruritic, erythematous eruption on her central chest area. She was otherwise in good health.

On examination, a dermal erythematous eruption with well-defined borders was observed on the anterior aspect of the chest (**Figure 1**). The eruption consisted of both macules and papules. A punch biopsy specimen from the erythema was stained with hematoxylin-eosin (**Figure 2**) and alcian blue (**Figure 3**).

What is your diagnosis?



Figure 1.

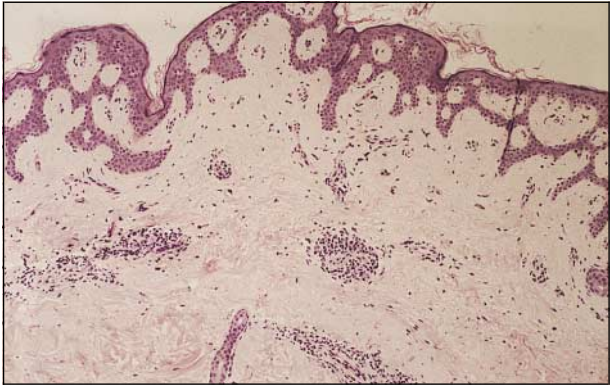


Figure 2.

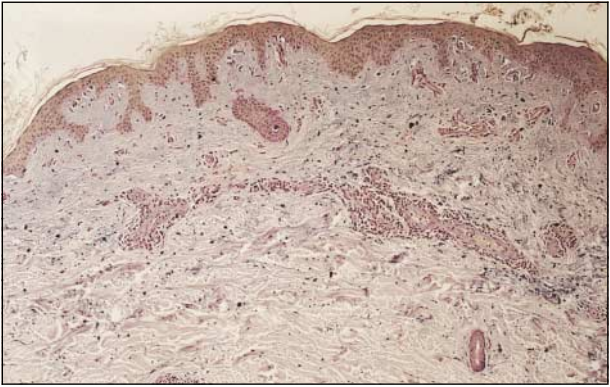


Figure 3.

Giant Tumor of the Back

Diagnosis: Giant amelanotic malignant melanoma.

MICROSCOPIC FINDINGS AND CLINICAL COURSE

On gross examination, the tumor was nodular, ulcerated, and at least 4 cm thick. Histologically, there was a well-developed junctional melanocytic proliferation. The melanocytes also showed confluent dermal growth, a lack of maturation, and prominent cytologic atypia, with anisonucleosis and large nucleoli. Abundant mitotic figures were present throughout the lesion. Several areas of identifiable melanin pigment were seen in the routine sections. Immunohistochemical staining was markedly and diffusely positive for S100 protein and HMB-45. In summary, these features were thought to be consistent with a primary melanoma.

The patient was admitted to the hospital surgical service for excision of the tumor. A wide-margin excision was performed down to fascia, and the surgical site was closed with a partial-thickness skin graft. The patient had an uneventful hospital stay and was discharged with an appointment for a positron emission tomographic scan and standard postoperative follow-up. Three weeks later, he began experiencing left leg weakness and an inability to sit up or pass urine. Within 3 days, he developed total paralysis of his lower limbs and placed a call to the emergency medical service. A magnetic resonance imaging scan of the spine revealed a single metastasis to the anterior T3-T4 vertebrae, with collapse of the anterior body of T3. Computed tomographic scans of the chest, abdomen, pelvis, and brain revealed innumerable metastases in the lung fields. No surgical procedure, radiation therapy, or che-

motherapy was performed. The patient was discharged home with hospice care.

DISCUSSION

Malignant melanomas presenting as a giant mass are very rare.¹ To our knowledge, this case represents the first report of a pedunculated, amelanotic melanoma presenting as a giant mass. Malignant melanomas of this size are almost never seen, because most patients seek medical attention at an earlier stage.¹

Amelanotic malignant melanomas make up between 2% and 8% of all malignant melanomas.² They have a predilection for sun-exposed areas but may occur anywhere on the body.² Peak incidence occurs in the sixth decade of life, and there is no sex predilection.² Even though amelanotic malignant melanomas tend to be clinically more advanced at diagnosis than their pigmented counterparts, the prognosis is the same for both types of melanomas after tumor thickness, location, sex, and patient age are adjusted for.³

Although amelanotic malignant melanomas may lack pigment clinically, it is often possible to demonstrate the pigment histologically.³ If special stains fail to reveal any pigment, immunohistochemical analysis using S100 protein, HMB-45, and Melan-A usually confirms the presence of melanocytes within the tumor.³ Amelanotic malignant melanomas occur in all clinical subtypes of cutaneous melanomas but most frequently as desmoplastic (50%) and subungual (25%) types.³ Absence of pigment often can make diagnosis of these tumors difficult, which may in part explain their more advanced stage at diagnosis. Amelanotic melanomas have been confused clinically with a variety of benign and malignant

Recurrent Nodules on the Feet of a Child

Diagnosis: Recurrent infantile digital fibromatosis.

MICROSCOPIC FINDINGS AND CLINICAL COURSE

The biopsy specimen revealed an intradermal benign neoplasm composed of spindle cells with blunt, vesicular nuclei arranged in interweaving fascicles. There were characteristic intracytoplasmic eosinophilic globules measuring approximately 5 to 9 µm in diameter.

The clinical and histologic findings led to a diagnosis of recurrent infantile digital fibromatosis. After being informed of the benign nature of these lesions, the patient's mother preferred conservative treatment with observation only.

DISCUSSION

Recurrent infantile digital fibromatosis is also known as inclusion body fibromatosis or Reye tumor.¹ In 1965, Reye² first described the staining characteristics of the pathognomonic intracytoplasmic paranuclear inclusion bodies that histologically distinguish this lesion from other benign fibromyofibroblastic lesions that occur during infancy and childhood. The list of differential diagnoses includes fibrous hamartoma, infantile myofibromatosis, infantile

desmoid-type fibromatosis, fibromatosis colli, calcifying aponeurotic fibroma, and hyaline fibromatosis.

Most patients present at birth or in the first year of life with asymptomatic smooth, pink or skin-colored nodules that are smaller than 1 cm in diameter. There is a predilection for the lateral or dorsal aspect of the fingers or toes, sparing the thumbs and great toes in all cases. The lesions may be solitary or multiple, involving the same or different digit. Males and females are affected equally. Inclusion body fibromatosis outside the digits has been reported in older children and adults on the extremities and tongue and within an endocervical polyp.^{3,4}

The histologic and electron microscopic characteristics of infantile digital fibromatosis include an intradermal neoplasm composed of interlacing bundles of myofibroblasts that exhibit a rich supply of granular endoplasmic reticulum and a prominent Golgi zone embedded within an abundant collagenous matrix.⁵ The 1.5- to 10-µm intracytoplasmic inclusion bodies, once regarded as viral in origin, are now considered to represent abnormal contractions of randomly raveled 5- to 7-nm actinlike filaments.⁵ The inclusions are continuous, with bundles of actin filament, and contain dense bodies and membrane-bound vesicles, the latter considered to be entrapped cell organelles.⁵ The inclusions stain pale pink with hematoxylin-eosin, bright red with Masson trichrome, yellow with van Gieson, and purple with phosphotungstic acid-hematoxylin.

Vulvar Lesion in a 45-Year-Old Woman

Diagnosis: Schistosomiasis of the vulva.

MICROSCOPIC FINDINGS AND CLINICAL COURSE

The vulvar biopsy specimen revealed a noncaseating granulomatous lesion composed of epithelioid histiocytes and giant cells surrounded by a thin rim of lymphocytes. Also, there were *Schistosoma* ova surrounded by granulomatous inflammation. The *Schistosoma* ova showed a terminal spine characteristic of *Schistosoma haematobium*. Ziehl-Neelsen, periodic acid-Schiff, and Fite stains were negative for organisms. There was no evidence of schistosomiasis elsewhere in the body.

Schistosomiasis was presumed to have developed 2 years before, when the patient was bathing in the Mali river in West Africa. The first lesion appeared 6 months later. The patient was treated with a single course of praziquantel (40 mg/kg), with the clearance of all lesions within 2 months. No relapse was observed after 2 years of follow-up.

DISCUSSION

Schistosomiasis is a chronic trematode infection that is endemic in many areas of the world and affects approximately 200 million individuals. Schistosomiasis due to *S haematobium* is endemic in Egypt and occurs in most parts of Africa and the Middle East.^{1,2} Infection develops after persons bathe or swim in stagnant infested water. Symptoms may not appear until months or years af-

ter exposure to the parasite. Usually, the disease produces complaints involving the urogenital tract, gastrointestinal tract, liver, or spleen. Dermatologic manifestations may occur with all 3 subspecies (*S haematobium*, *Schistosoma mansoni*, and *Schistosoma japonicum*) at all different stages of the schistosome life cycle.^{3,4}

Three different dermatologic manifestations of schistosomiasis are common.⁴ Swimming or bathing in infested waters can lead to infection by active penetration of the intact skin by cercariae emitted by the snail intermediate host (*Bulinus* for *S haematobium*). At this stage, there is a local allergic reaction to the cercaria after penetration of the skin, which is called *cercarial dermatitis* or *swimmer's itch*. The allergic reaction is characterized by a pruritic papular eruption that may occur a few hours after penetration of the skin and is due to irritation by the cercariae themselves.

Four to 6 weeks after infection, an urticarial reaction may occur along with fever, edema, headache, arthralgia, abdominal pain, and hypereosinophilia, which is believed to be a hypersensitivity response, possibly immune complex mediated, to the presence of the parasite.⁵

The third cutaneous manifestation is known as *bilharziasis cutanea tarda*. After entering the body, the parasite migrates through the lungs and liver into the portal venous system. After leaving the portal circulation and reaching the inferior mesenteric veins, adult flukes lodge in the pelvic venous plexi.⁵ There, they produce ova that become trapped in the tissue, triggering a granulomatous inflammatory response. Patients can develop genital or perianal wartlike lesions and, rarely, ectopic cuta-

Erythematous Rash on the Chest

Diagnosis: Reticular erythematous mucinosis (REM) syndrome.

MICROSCOPIC FINDINGS

The hematoxylin-eosin-stained biopsy specimen revealed a normal epidermis and a deep perivascular and periadnexal infiltrate of lymphocytes. Alcian blue staining showed the separation of the collagen fibers by mucin.

DISCUSSION

In 1960, Perry et al¹ reported a unique type of mucinosis with a predilection for the thorax and gave it the name *plaque-like mucinosis*. In the early 1970s, Steigleder et al² reported 4 cases with a sheetlike or netlike erythema that affected the upper part of the chest or back. They found no evidence of lupus erythematosus, metabolic disease, or hematologic disease. All 4 patients responded to treatment with antimalarial agents. Steigleder and colleagues called this condition *REM syndrome*. It is now believed that REM syndrome and plaque-like cutaneous mucinosis are both part of the same disease spectrum.³

REM syndrome is rare and usually affects young adult or middle-aged women but has been reported in children.⁴ The eruption classically involves the central part

of the chest and upper back area and is irregular but with a well-defined border and areas of pink reticulate macular erythema. A papular component has been noted in some cases. The papular component may be prominent at the periphery, resulting in a raised palpable border. There is usually no pruritus, although itching may occur after sun exposure and the erythema may become more apparent.⁵ The erythematous areas become more infiltrated with time and slowly increase in size.

Histologically, REM syndrome has a characteristic picture. The epidermis is normal, and a mild to moderate perivascular lymphocytic dermal infiltrate is seen. Alcian blue staining may be used to demonstrate mucin deposits. Bleehan et al⁶ noted that colloidal iron staining was more sensitive than alcian blue in demonstrating acid mucopolysaccharides. The results of direct immunofluorescence are usually negative.⁶ Topical steroid therapy tends to be unhelpful in controlling the eruption, although antimalarial agents are usually effective.^{7,8} Our case responded to hydroxychloroquine sulfate at a dosage of 200 mg twice daily. Cyclosporine has been shown to be ineffective in the treatment of this condition.⁹

Lupus erythematosus may resemble REM syndrome. Both conditions demonstrate a female preponderance, photoexacerbation, and clinical response to antimalarial agents. Antinuclear antibodies and antibodies to the cytoplasmic antigens Ro (SS-A) and La (SS-B)

disorders, such as intradermal nevus, seborrheic keratosis, verruca vulgaris, dermatitis, pyogenic granuloma, nevus depigmentosus, granuloma annulare, scar, actinic keratoses, lymphocytoma cutis, basal cell carcinoma, keratoacanthoma, Bowen disease, Merkel cell carcinoma, atypical fibroxanthoma, and cutaneous metastases from internal malignancies.^{3,4}

Malignant melanoma is now the fifth most common cause of cancer in the United States,⁵ and the incidence has increased faster than that of any other cancer in this country. Malignant melanoma presents at an earlier age than most other cancers, and the individuals who die of malignant melanoma also tend to do so at an earlier age.⁵ Melanoma is one of the leading cancers in terms of average years lost per person.⁵ Since 5-year survival for clinical stage I melanoma is directly related to tumor thickness, diagnosing these tumors while they are still relatively thin is essential.⁶

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The nodules can rarely cause contractures or deformity.⁶ Regression can occur spontaneously or after partial resection, usually within 4 years.⁷ Local recurrence at the excision site, however, happens more than 60% of the time, most often within several months. Recurrence is thought to occur as a result of incomplete excision,⁸ although recurrence after histologically confirmed free margins has also been described.⁹ Surgical treatment is recommended only in cases in which there is functional impairment or aggressive growth.

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neous granulomatous reactions on the trunk.⁶ Bilharziasis cutanea tarda seems to represent 0.2% of the bilharziosis manifestations⁷ and occurs with *S haematobium* in 70% of cases. It usually affects the genital area, most frequently the vulva. In most cases, the lesions were found on the labia majora,⁷ possibly as a result of the rural practice of circumcision in which the labia minora are removed.

Praziquantel is the antihelminthic drug of choice, giving high cure rates for cutaneous and systemic infections caused by all species of *Schistosoma*. Normally, a single 40-mg dose of praziquantel is sufficient for cure and is well tolerated by the patient. Lesions heal a few weeks after administration.⁸

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should be looked for in all REM cases. REM syndrome has been reported in association with both hyperthyroidism and hypothyroidism, discoid lupus erythematosus, carcinoma of the breast and colon, and thrombocytopenic purpura.⁷

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