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Sebaceous carcinoma on the arm of a 10-year-old girl

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Abstract

We report a case of a 10 year-old girl diagnosed with sebaceous carcinoma of the posterior left arm. The presented case reviews the histopathological and immunohistochemical characteristics of this malignancy, including a review of the literature in pediatric patients regarding prognosis and treatment. Sebaceous carcinoma is a malignant neoplasm with sebaceous differentiation, typically occurring in the sixth-to-seventh decades of life. It most commonly arises in the periocular region. It is extremely rare in the pediatric population.

Keywords: dermatopathology; papules; malignant neoplasms; sebaceous carcinoma

Case Synopsis

A 10-year-old girl was referred to the dermatology clinic for evaluation of a tender lesion that had been slowly growing for one year. The lesion was a solitary 8mm round pink papule with a collarette of scale on her left posterior arm that was painful to manipulation (**Figure 1**). The papule was initially diagnosed as a boil and was unsuccessfully treated with warm compresses, antibiotics, and incision and drainage. Past medical history was notable only for seasonal allergies, for which she took occasional antihistamines. The patient took no other medications. Family history could not be established as the patient had been adopted from China at the age of ten months.

Biopsy of the papule revealed numerous sebocytes with sheets and nodules of infiltrative basaloid epithelial cells. Also observed were focal neutrophilic



Figure 1. Firm, tender 8 mm dome-shaped pink papule with collarette of scale on the left dorsal arm.

microabscesses, areas of necrosis, holocrine disintegration in the center of the tumor lobules, and a fibroblastic stroma extending to the deep dermis. Tumor cells showed atypia, high mitotic activity, and clusters of cells with extensive sebaceous differentiation (**Figures 2-5**). No pagetoid spread was noted within the epidermis. Immunohistochemical stains showed diffuse membranous and cytoplasmic reactivity with antibodies to epithelial membrane antigen (EMA).

The patient was diagnosed with sebaceous carcinoma. Further immunohistochemical staining for MutL Homolog 1 (MLH-1), MutS Homolog 2 (MSH-2), and MutS Homolog 6 (MSH-6) showed normal retained nuclear staining within the tumor cell population. These results argue against Muir-Torre syndrome as a cause for our patient's tumor. However, further microsatellite instability studies were not carried out. The tumor was excised with clear margins and the patient is doing well without

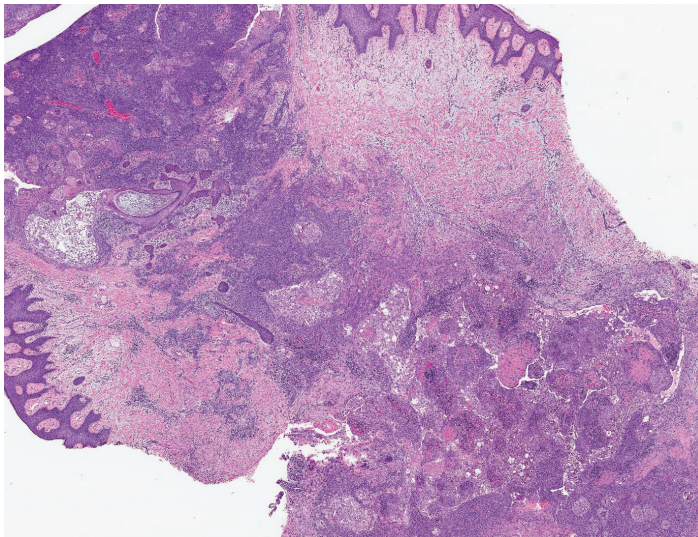


Figure 2. Low-power photomicrograph stained with H&E, 4x. Sheets and nodules of infiltrative basaloïd germinative cells, with areas of squamous differentiation, necrosis, and focal neutrophilic microabscesses.

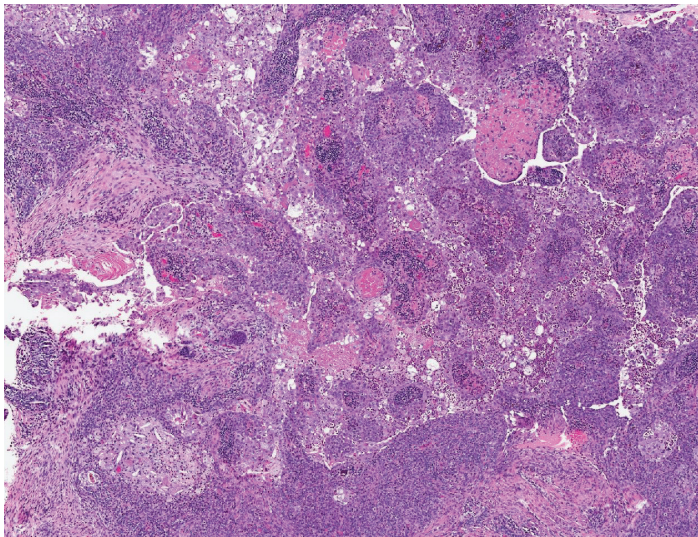


Figure 3. Medium-power photomicrograph stained with H&E, 10x. Variably hypercellular fibromixoid stroma surrounding and enveloping tumor cells with focal mucinous and cystic changes in portions of the tumor.

recurrence or metastatic disease after a follow-up of three years.

Case Discussion

Sebaceous carcinoma is a rare and potentially aggressive skin cancer arising from sebaceous glands. Although it may originate wherever sebaceous glands are found in the body, about 75% of cases develop in the periocular region. It represents approximately 3.2% of all eyelid malignancies and 0.8% of all eyelid tumors [1]. It shows a high tendency towards metastasis and local recurrence. It is more common in the seventh and eighth decades of life, but it may

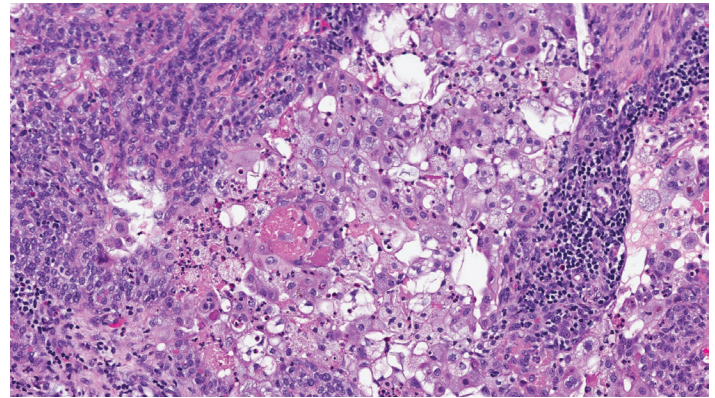


Figure 4. High-power photomicrograph stained with H&E, 40x. Extensive sebaceous differentiation in tumor cells throughout the lesion with both discrete sebaceous nodules and foci of apoptosis and necrosis within the tumor island. Tumor cells show areas of prominent mitotic activity.

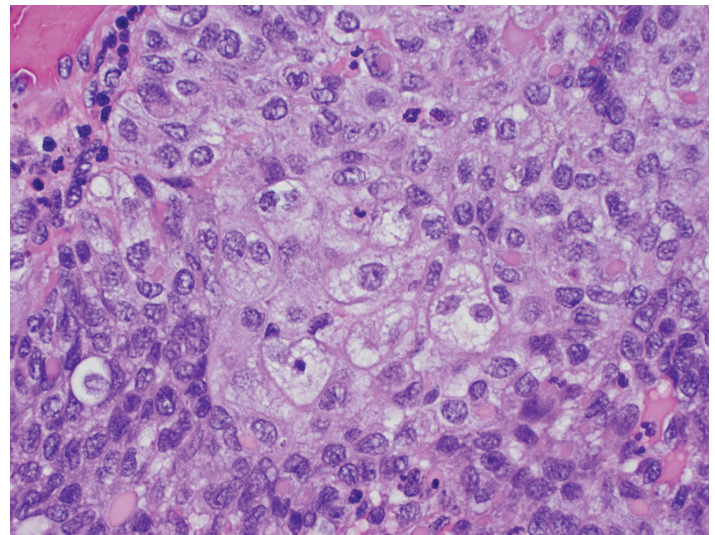


Figure 5. High-power photomicrograph stained with H&E, 100x. Scattered atypical mitotic figures, necrotic tumor cells, and typical sebaceous differentiation with vacuolated cytoplasm and scalloped nuclear borders are noted.

occur at any age [2]. It is extremely rare in children with approximately ten cases having been reported in the literature [1]. There is consequently a paucity of evidence to guide management of sebaceous carcinoma in pediatric patients; evidence regarding diagnosis and treatment is derived primarily from the adult population.

The cause of sebaceous carcinoma is usually unknown. It may be associated with Muir-Torre syndrome (MTS), an autosomal dominant condition caused by a defective mismatch repair gene that results in microsatellite instability. MTS is

characterized by at least one sebaceous neoplasm paired with at least one visceral malignancy, with colorectal carcinoma being the most common [3]. Most cases are not associated with MTS and a link between MTS and pediatric sebaceous carcinoma has not been demonstrated [1].

Sebaceous carcinoma typically presents on the eyelid as a slowly-growing, firm, red or yellow nodule. Extraocular lesions account for about 25% of cases and present variably as a pink, yellow, or red nodules of varying size that may show ulceration. Diagnosis is often delayed for years owing to the lesion being misdiagnosed as a chalazion or hordeolum [2, 4]. These tumors may grossly mimic basal cell carcinoma, squamous cell carcinoma, or cutaneous horn. Advanced sebaceous carcinoma may spread to the conjunctiva or lower eyelid and may result in ulceration, eyelash loss, vision loss, and metastasis.

Suspicious lesions should be evaluated with a full-thickness biopsy as soon as sebaceous carcinoma is suspected. Some sources have recommended establishing the diagnosis with fine needle aspiration. However, the sensitivity has not been established and equivocal findings must be confirmed with full-thickness biopsy [4].

Histologic findings of sebaceous carcinoma include mixed basaloid and squamoid cells infiltrating the dermis in bands and nodules with tumor cells expressing sebaceous differentiation or vacuolization. Tumor cells may show pagetoid spread, extending to the adjacent epithelia beyond the main tumor. This may occur in the conjunctivae, cornea, or adjacent skin [1, 4]. Poorly-differentiated tumors may mimic basal cell carcinoma. Positive immunohistochemical staining for epithelial membrane antigen (EMA), adipophilin, and androgen receptor may be helpful in confirming sebaceous differentiation [1].

The preferred treatment is surgical excision with adequate margins [1, 4]. Mohs surgery has shown improved remission rates when compared to traditional excision, which shows up to a 30% rate of recurrence. Radiation has been used successfully in a few cases and may be considered if excision is contraindicated [5].

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