CYTOLOGIC CHARACTERISTICS OF A MALIGNANT PROLIFERATING TRICHILEMMAL CYST

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Abstract : The proliferating trichilemmal cyst (PTC) is a tumor that originates from hair follicles. It is almost always benign, but rare lesions have been reported with malignant potential. In the present case, the patient had a mass on the right side of her neck for a few years, which suddenly increased in size. Cytologic examination revealed many atypical squamous cells staining well with orange G and light green, suggestive of a squamous cell carcinoma. Total extirpation was performed under general anesthesia, gross pathology revealing a focally cystic tumor approximately 5 cm in diameter. The final diagnosis was malignant PTC from the characteristic features. We here report cytologic findings for this case with regional lymph node metastasis.

Key words: proliferating trichilemmal cyst, squamous cell carcinoma, metastasis, cytology

INTRODUCTION

The proliferating trichilemmal cyst (PTC), also called the proliferating trichilemmal tumor (PTT)[1], is an uncommon appendageal skin neoplasia originating from the outer root sheath of the hair follicle. It generally occurs as a single lesion, mainly on the scalp in elderly women[2]. Histologically, it exhibits variably sized lobules composed of atypical squamous epithelium, characteristically showing trichilemmal keratinization, an absence of keratohyaline granules and abrupt change into eosinophilic amorphous keratin in the centers of the lobules. Some of these latter are surrounded by a vitreous layer and show palisading of their peripheral cell layer[1]. There is usually a slight degree of nuclear anaplasia, and some tumors show vacuolization or clear cell formation as a result of glycogen storage[3].

PTC is usually benign, but in very rare instances, local recurrence and metastasis have been reported. There have been 14 case reports of malignant PTC so far, to our knowledge. The characteristic histological features are severe dysplasia of squamous epithelium and occasional invasion into the surrounding tissue[4].

Reports, particularly of malignant lesions, are rare because primary tumors arising from epidermal appendages are commonly diagnosed by excisional histology[5].

We experienced a case of malignant PTC with regional lymph node metastasis and the opportunity to obtain detailed cytological findings. We describe these and review the literature on this type of tumor.

CASE REPORT

A 56-year-old woman visited the hospital for a solitary subcutaneous mass which had been present on the right side of her neck for a few years. The tumor had rapidly increased in size since one month previously and a metastatic cancer of the neck lymph node was therefore clinically suspected. A biopsy from the lesion was carried out. At the same time, imprinting cytology was performed. Although a metastatic squamous cell carcinoma (SCC) was strongly suspected, there were no gross findings indicative of a primary tumor in the pharynx, tonsils, esophagus, or stomach by endoscopic examination. Therefore, total extirpation under general anesthesia was performed and a focally cystic tumor, approximately 5 cm in diameter, was obtained.

CYTOLOGICAL AND HISTOLOGICAL FINDINGS

Imprinting cytology was performed at the time of biopsy. Cytologically, many atypical

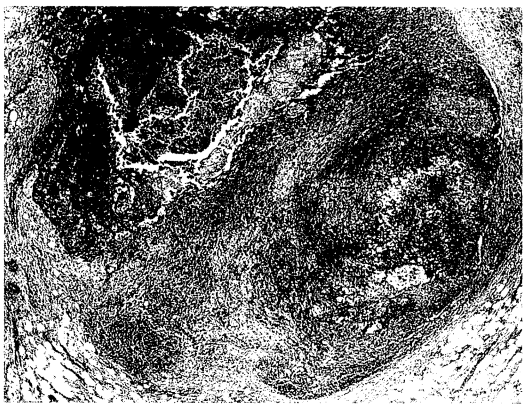

Fig. 1. Atypical keratinizing and nonkeratinizing squamous cells staining well with orange G and light green, evident against a background of slight inflammation.

Fig. 2. Small atypical cells staining well with light green partially proliferating in sheets.y

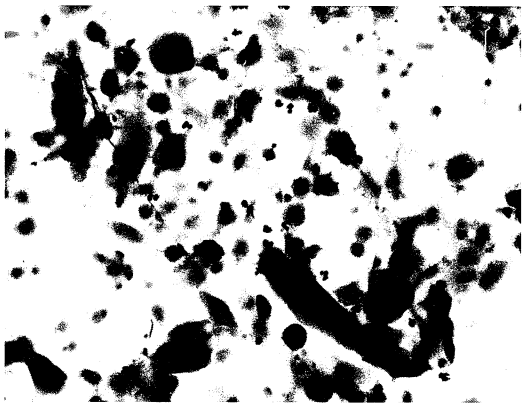

Fig. 3. The tumor exhibits central cystic change. Note the atypical squamous cells proliferating in the cyst wall.

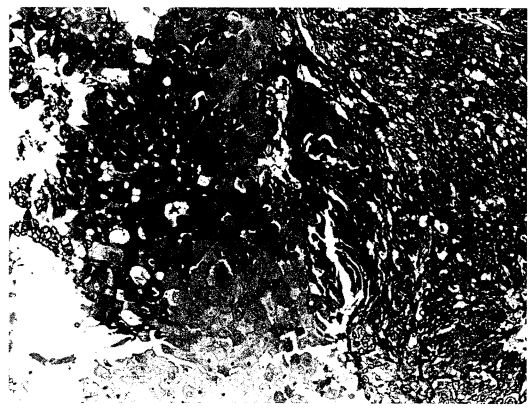

Fig. 4. Atypical squamous cells, proliferating in a regional lymph node.

keratinizing and nonkeratinizing squamous cells with abundant cytoplasm staining well with orange G and light green were recognized against a background of slight inflammation (Fig. 1). Individual cells tended to be isolated, small atypical cells staining well with light green partially proliferating in sheets (Fig. 2). Both keratinizing and nonkeratinizing squamous cells exhibited various sizes and shapes, for example, oval, polygonal, and sometimes elongated. Some oval atypical cells showed a concentric circle pattern in their cytoplasm. Nuclear pleomorphism was also seen, and disappearance of nuclei and pyknosis were comparatively frequent, associated with keratinization or necrosis.

Histologically, the biopsy specimen showed atypical eosinophilic squamous cells, and a metastatic SCC was suspected. After surgery, the totally extirpated tumor was found to show central cystic change, the cyst walls consisting of basal proliferating basophilic squamous cells with moderate nuclear atypia and no apparent cytoplasmic keratin, and more superficial eosinophilic squamous cells with mild nuclear atypia and prominent cytoplasmic vitreous material with few granular cells (Fig. 3). The large part of tumor in the cyst showed carcinomatous change and the tumor focally invaded into the capsule. Foreign body giant cells were also apparent. Moreover, similar findings were recognized in a regional lymph node (Fig. 4). Vascular invasion was not found in this specimen.

Pathological diagnosis: Malignant proliferating trichilemmal cyst

DISCUSSION

Follicular cysts that originate from hair follicles are classified into two types. The more common is the epidermal type, usually called the epidermoid cyst, and the other is the so-called pilar or trichilemmal type. With the latter, cysts without a granular layer occur, predominantly on the scalp. McGavran et al. proposed they be called pilar cysts for the first time in 1966, because of their histological similarities to outer root sheaths of hair follicles under the light and electron microscopes[6]. The cysts occasionally exhibit some degree of hyperplasia of the lining epithelium; Pinkus named this type of tumor the PTC in 1981. All types of transitional forms can be found between the common trichilemmal cyst and full-blown PTC and with regard to their histogenetic relations, Browstein and Arluk proposed that trauma and inflammation might induce a trichilemmal cyst to proliferate and show a broad spectrum of cellular atypia of pseudocarcinomatous proportions[2].

With regard to actual malignancy, Sau et al. reported ten lesions demonstrating carcinomatous change, including one anaplastic type, with regional lymph node metastasis[7]. Weiss et al. described a case of a 78-year-old man featuring multiple common cysts and two PTCs, one of which showed malignant transformation as evidenced by metastasis[8]. Recently, Park et al. reported a round mass in the occiput of a 32-year-old man recurring twice after wide excision, which eventually metastasized to the cervical lymph nodes, periparotid area, and chest[9].

Histologically, both PTC and malignant PTC show atypical squamous cells to some extent, and tend to be misdiagnosed as SCC. In differentiating PTC or malignant PTC from SCC, abrupt delineation of areas of keratinization, low mitotic activity, minimal pleomorphism, and sharp demarcation between the stroma and adjacent dermis are helpfu[19]. Moreover, Noto et al. reported that differential diagnosis of benign and malignant PTC could be made on the

basis of mitotic rate, cytological and architectural atypia, necrosis, and stromal infiltration. In the present case, not only trichilemmal keratinization and mild to moderate nuclear atypia but also invasion of surrounding tissue and metastasis to a regional lymph node were recognized.

While the clinical value of cytology in dermatology is well-known, most primary tumors arising from epidermal appendages are diagnosed by excisional histology[5]. There are a few reports of fine needle aspiration biopsies of trichilemmal cysts or PTC, Slater et al. describing the former to aspirate easily and show amorphous debris with birefringent crystalline material[10]. Regarding PTC, Biernat and Kordek reported small squamoid cells to be predominant in smears, occurring mainly in clusters and sheets without prominent atypia and complete keratinization[11]. It is predictable that loss of the granular layer could not be identified by cytology, even if the structures of the tissue were clear on imprinting. Therefore it is difficult to distinguish malignant PTC from SCC by cytological examination only, if amorphous debris equivalent to keratin is not recognized.

With our imprinting cytological examination, a number of moderately atypical keratinizing and nonkeratinizing squamous cells were seen, reflecting the histological picture. While amorphous debris was not obviously recognized in the specimen at that time, the tumor was diagnosed as SCC, and later the malignant PTC was identified only on histological examination.

Recently, it has been proposed that the malignant PTC is in fact a type of SCC, Ackerman et al. coining the term proliferating trichilemmal cystic squamous cellcarcinoma (PTCSCC)[2]. Containing the origin it is not fully understood, and differences in prognosis between malignant PTC and SCC derived from epidermis remain to be reported.

Whatever the case, when a cyst or tumor is diagnosed in dermis by cytology, it is important to bear in mind the possibility of malignant PTC and concentrate attention on differentiation from SCC.

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