

Atypical Proliferating Trichilemmal Cyst in Breast.

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ABSTRACT

Trichilemmal or Pillar cyst is one such entity, which presents in both a non-neoplastic and neoplastic form. These cysts occur preferentially in areas with dense hair follicle concentration, therefore 90% occur in the scalp. Moreover, they are the most common cutaneous cysts in the scalp and secondly common in the head and neck region. Pillar cysts are almost always benign, with malignant transformation occurring rarely. In 2% of cases, pillar cysts occur either single or multiple[1].We present a case encountered at NCI-Sabratha with literature review.

Keywords: Malignant Proliferating Trichilemmaltumor; Proliferating Trichilemmal tumor; Squamouscell carcinoma; Pilar tumor;Epidermoid cyst.

Abbreviation:

National Cancer Institute (NCI), Proliferating TrichilemmalTumors (PTT), Malignant Proliferating Trichilemmal Tumor (MPTT), TrueCut Biopsy (T.C.B), Squamous Cell Carcinoma (SCC), Breast Image Radiology and Data Base (BIRADs).

INTRODUCTION

Proliferating Trichilemmal Tumors (PTT), also known as proliferating pillar tumors, are rare neoplasms of the external root hair sheath that are largely benign, cystic in nature, and characterized as containing trichilemmal keratin. These are rare exophytic tumors and mainly confined to the scalp and back of the neck, and most often

reported in middle age females [2]. The potential for PTT to undergo transformation to MPTT is unknown, with only about 40 cases of MPTT having ever been reported in the English literature.

We present an exceedingly rare case of a 34-years-old female with an 2X2 CM mass of the left breast, which was initially diagnosed as squamous cell carcinoma and later

classified as proliferating trichilemmal tumor.

Case presentation

A 34-years-old Libyan woman was evaluated in our institute for recent onset of pain and swelling of left breast since one year back. She denied breast irritation, prolonged skin disease, trauma and oral contraceptives use in the past. She gave birth to four children and had no family history of breast cancer. Her general examination was unremarkable.

Locally revealed a palpable well circumscribed and firm mass, measuring about 2x1 cm, present in the inner upper quadrant of her left breast. The right breast examination was normal. There was no palpable lymph nodes, including the axillary and supraclavicular regions.

An ultrasound scan of the breast showed hypoechogenic solid lesion about 2x2.5 cm seen at 9 o'clock on left breast without significant lymph nodes seen bilateral category (BIRADs II).

T.C.B was done, and histopathology report illustrated squamous cell carcinoma. Metastasis was excluded by total body CT scan. Subsequently, wide local

excision of the left breast with ipsilateral axillary lymph nodes dissection were performed, which were free of tumor.

Routine histopathologic analysis of the specimen demonstrated a proliferation of squamous cells in which there were extensive areas of amorphous keratinization [Figure 1]. This finding is in line with the follicular differentiation. Therefore a diagnosis of a proliferating trichilemmal tumor rather than a cutaneous squamous cell carcinoma was made.

A bundle of eosinophilic keratin was seen in center of lobules. It is the same type of ordinary hair follicle keratinization mimic of squamous cell carcinoma, but, no frank atypia or abundant mitosis or necrosis [Figure 2]. Though, p53 [Figure 3] and KI67 (3%) [Figure 4] can be increased even in proliferating trichilemmal cyst and not necessarily specific for SCC. In MPTT usually there is loss of CD34 immunostain which is another clue in border line cases.

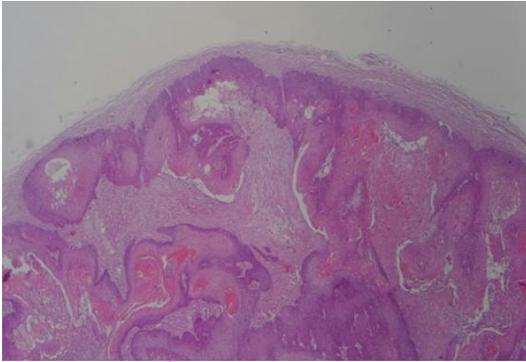


Figure 1: Cystic tumor portion where keratin amorphous deposits are observed (H and E, ×10).

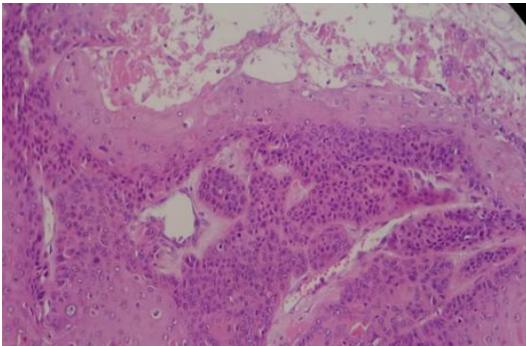


Figure 2: Typical single lesion non infiltration.



Figure 3: p53 Immunostain

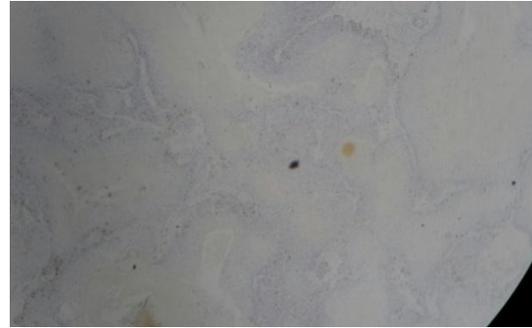


Figure 4: KI67 Immunostain

Discussion

PTT of the scalp are relatively uncommon and of the chest is an even rarer condition that has been infrequently reported. These tumors were first named as proliferating epidermoid cysts by Wilson Jones in 1966 [2]. In our institute since 2006 till 2014, 1883 patients were seen only this case was encountered. It was misinterpreted both clinically and histologically as squamous cell carcinoma. PTT five times more likely to occur in females and have a median age of occurrence of 65-year-old [3].

Areas devoid of non-terminal hair, such as lanugo hair follicles of the bald scalp are unlikely to produce these tumors [4, 5]. Pillar tumors are more common in areas with excess hair growth [4, 5] and less common in areas such as the anterior chest of a female as in our case, or of the skin over the breast as reported by Uchida et al [6]. The differential diagnoses of malignant

proliferating trichilemmal tumors includes; squamous cell carcinoma, trichilemmal carcinoma, proliferating trichilemmal tumors, sebaceous cyst, and angiosarcoma. So, It is essential to properly diagnose MPTTs due to differences in aggressiveness and treatment course.

The current recommended management of MPTT is local excision with margins of normal tissue and a work-up to exclude metastases along with routine long-term follow-up [6]. Our case not only demonstrates the difficulty and importance of making a proper diagnosis, but also adds to the literature of these rare tumors. In theunluckily patient, who does not follow the typical presentation of a MPTT as noted in previously reported cases.To our knowledge this patient represents the youngest reported case of PTT.

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