# Pilomatricoma masquerading as a parotid mass in a child-a diagnostic challenge on cytology

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#### **Abstract**

Pilomatrixoma is a slow growing benign skin neoplasm of hair matrix origin. This tumor is common in children and young adults, especially in the head and neck region. Because of its varied morphological spectrum on cytology it can pose diagnostic difficulties and cause unnecessary treatment. We report a case of 5 year old girl who presented with a nodule over right parotid region. Fine needle aspiration cytology was done which was suspicious of mucoepidermoid carcinoma; however, histopathological diagnosis was Pilomatrixoma. On review of literature we observed that pilomatricomas have been quite often misdiagnosed especially on cytology. Our case highlights the diagnostic pitfalls of pilomatricoma on cytology.

Key-words: Pilomatricoma, Cytology, Parotid region

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#### Introduction

Pilomatricoma or *calcifying epithelioma of Malherbe* is a benign skin tumor of hair matrix differentiation [1]. In children it is commonly seen in the head and neck region and is more frequent in girls than boys [2,3]. It is asymptomatic, deep seated, firm, subcutaneous mass adherent to the skin, ranging from 0.5-3 cm in size and rarely as large as 5 cm [4]. On histopathology it is

easily diagnosed because of its classic morphological features but on FNAC it often gives rise to diagnostic difficulties owing to wide cytomorphological spectrum. Fine-needle aspiration cytology plays an important role in the pre-operative diagnosis. Exact pre-operative cytological diagnosis helps in the management of the case, as surgical excision is curative.

# **Case History**

A 5 year old girl child presented with a swelling in the right parotid region since 3 months. On examination a 1 x 2 cm firm mobile nodule with smooth surface was noted. No cervical lymphadenopathy was seen. Other systemic examination was normal. Lab investigations such as complete haemogram, Serum biochemistry, chest X-ray & ultrasonography of abdomen were normal. A clinical diagnosis of right parotid tumor was made and the case was referred to cytopathology department for FNAC of the nodule. Aspiration was done in the cytopathology section using 23 gauge needle and syringe. The aspirate was abundant and whitish. Smears were made and fixed in 95% alcohol and stained by Papanicolaou (PAP) and May Grunwald Giemsa (MGG) stain. Smears were cellular, showed predominantly cells arranged in cohesive clusters & dispersed singly [Figure 1 shows high cellularity, 4x PAP stain]. Cells showed indistinct cytoplasmic borders with moderate to scant amount of basophilic cytoplasm [Figure 2 shows cytoplasmic features, 40x MGG stain]. Nuclei had granular / fine chromatin with few of them showing prominent nucleoli [Figure 3 shows nuclear features, 40x PAP stain]. Also noted were squamoid cells, giant cells, macrophages & keratin [Figure 4 shows squamoid areas with keratin, 20x PAP stain]. Based on these features a diagnosis of parotid tumor, possibly a mucopeidermoid carcinoma was made.

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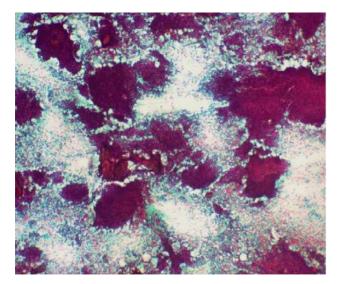


Figure 1: Shows high cellularity, 4x PAP stain

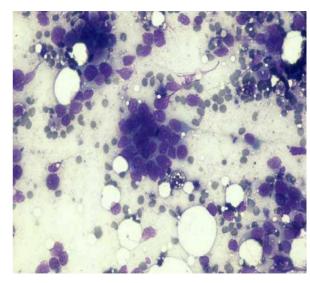


Figure 2: Shows cytoplasmic features, 40x MGG stain.

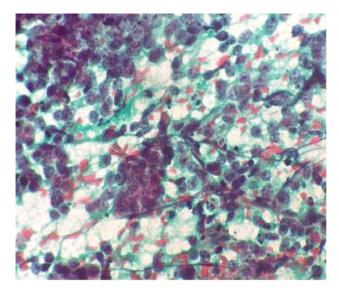


Figure 3: Shows nuclear features, 40x PAP stain

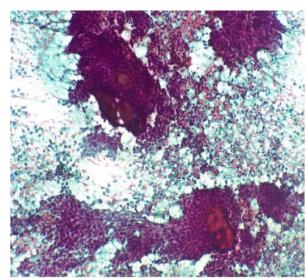


Figure 4: Shows squamoid areas with keratin, 20x PAP stain.

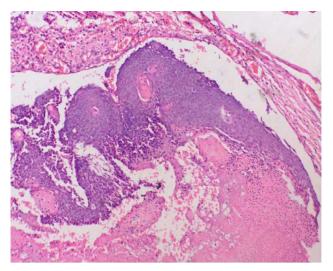


Figure 5: Shows basaloid cells & ghost cells, 10x hematoxylin and eosin stain

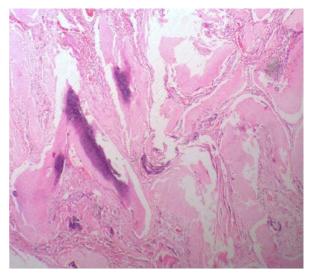


Figure 6: Shows ghost cells & calcifications, 10x hematoxylin and eosin stain

Subsequently a right total parotidectomy was done. Grossly specimen showed a well circumscribed nodule measuring 2.5x1.8x1cm. Cut surface was grey white with areas of calcification. Adjacent parotid gland was grossly unremarkable. Microscopy revealed well circumscribed subcutaneous lesion composed of islands of squamous epithelial cells made up of basophilic cells having hyperchromatic nuclei with little or no cytoplasm [Figure 5 shows basaloid cells & ghost cells, 10x hematoxylin and eosin stain]. Centrally the islands showed shadow cells having distinct cell border with unstained areas as shadows of lost nuclei. Inflammatory cells, foreign body giant cells & calcium deposits were seen [Figure 6 shows ghost cells & calcifications, 10x hematoxylin and eosin stain]. Based on these features a histopathological diagnosis of Pilomatricoma of parotid region was given. FNAC smears were reviewed because of the discrepancy; the predominant cell population which was thought as intermediate cells of mucopeidermoid carcinoma on cytology was in fact basaloid cells of pilomatricoma. Moreover, absence of ghost cells on cytology also deceived us.

#### Discussion

Pilomatricoma displays a spectrum of histopathologic features reflecting primarily different stages of growth. Histological features of pilomatrixoma show two types of epithelial cells: small basaloid cells and eosinophilic ghost cells without nuclei [5].

Early and well-developed lesions are characterized by small to large-sized, cystic lesions lined focally by aggregations of basaloid cells and few squamoid cells and filled centrally with large masses of eosinophilic cornified material containing ghost cells. Regressing lesions are relatively large cystic tumors with prominent areas of ghost cells and foci of basaloid and/or squamoid cells surrounded by a variable, inflammatory infiltrate [5]. Likewise on cytology also one can expect a varied cytomorphological spectrum depending on the stage of the growth. Because of this varied presentation many authors have misdiagnosed pilomatricomas on cytology. On cytological review of literature we found that Pilomatricomas on FNACs are usually over diagnosed as malignant lesions [6, 7].

Similarly in our case, because of cellular clusters of two cell population, dirty background, and absence of ghost cells on cytology, location of the lesion, rapid growth and young age we were suspicious of mucoepidermiod carcinoma of parotid. On review of cytological literature we came to know that in the parotid region, it may be mistaken for a mucoepidermoid tumor on cytology [8]. Likewise Luciano et al in his case report has mentioned that, pilomatricoma was misdiagnosed on cytology as mucoepidermoid carcinoma because of the basaloid cells of pilomatricoma were mistaken for intermediate cells of mucoepidermoid carcinoma [9].

Shivkumar S et al also had diagnostic difficulty on FNA which led to the three differential diagnosis of Mucopeidermiod carcinoma, Squamous cell carcinoma, calcifying odontogenic tumor [10].

The cytomorphological features of pilomatricoma are variable as documented in the literature. Our case delineates the cytomorphologic features of pilomatricoma mimicking carcinoma. Recognition of the unique constellation of cytological features in FNAC smears in the appropriate clinical context is most helpful in making this distinction.

Otherwise one can keep a possibility of differential diagnosis of Pilomatricoma on FNAC before labeling as malignant parotid tumor especially in children in whom this tumor is common. Aspiration from multiple sites may be contributory avoiding sampling error.

Pilomatricoma is often misdiagnosed entity in clinical practice also [11]. So correlation between clinical presentation and cytological findings are absolute prerequisites in arriving at the exact diagnosis thus avoiding unnecessary overtreatment. In conclusion, a cytopathologist should be aware of this varied array of appearance by pilomatricoma on cytology while dealing with head and neck tumors especially in children.

**What's known:** Pilomatricoma is a benign skin neoplasm. On histopathology it is easily diagnosed because of its classic morphological features and excision is the treatment of choice.

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