

Aggressive pilomatrixoma of the infra-auricular area: A case report

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Abstract

Although pilomatrixomas are well known among dermatologists and dermatopathologists, head and neck surgeons confronted with these lesions in the infra-auricular region do not consider this benign neoplasm in the differential diagnosis. Aggressive pilomatrixoma is a benign tumor of the hair matrix cells affecting mainly children. Histologically, the border between aggressive pilomatrixoma and pilomatrix carcinoma is still not clear. We report the case of a 15-year-old Turkish boy suffering from an aggressive pilomatrixoma of the infra-auricular region and review the literature about this unclear entity.

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1. Introduction

Pilomatrixoma is a benign tumor of the hair matrix cells that appears most frequently in children [1]. Most are benign and slow-growing and do not recur after excision. Pilomatrixoma is characterized histologically by dermal or subcutaneous aggregates of basaloid cells. A small number of aggressive or malignant variants have been reported. Those tumors could recur if not widely excised [2].

2. Case report

A 15-year-old Turkish boy presented with a 10-month history of a slowly growing left cervical mass. Examination of the head and neck area revealed a 2 cm × 2 cm subcutaneous irregular and mobile mass in the infra-auricular region. The cranial nerves were intact and

symmetrical. The rest of the ENT examination was normal and no cervical adenopathy was noted.

Computed tomography (CT) with intravenous contrast injection was performed. This procedure revealed a well-circumscribed mass in the infra-auricular region (Fig. 1) that neither showed evidence of loco-regional extension, i.e. involvement of the parotid gland or the parapharyngeal space, nor any signs of bony erosion. A fine-needle aspiration cytology revealed a highly cellularized material with a large number of round basaloid cells featuring a high nucleocytoplasmic ratio and a large number of mitotic figures (Fig. 2). Those histological features lead the pathologist to propose the disturbing diagnosis of small round cell tumor. This infra-auricular mass was therefore excised using a transcervical approach. Macroscopic examination of the tumor revealed a rather well circumscribed lesion showing however some capsular ruptures. The histological features are illustrated in Figs. 3 and 4. The lesion presented irregular borders and was composed of solid aggregations of basaloid cells surrounded by fibrous stroma. A high number of cellular atypias and a prominent mitotic activity were observed in those basaloid cells

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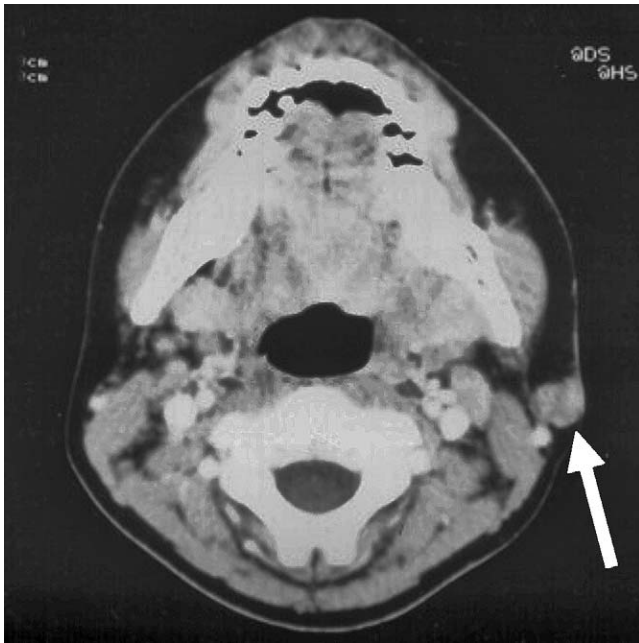


Fig. 1. Computed tomography (CT) with intravenous contrast injection revealed a well-circumscribed mass in the infra-auricular region (arrow) that neither showed evidence of loco-regional extension, i.e. involvement of the parotid gland or the parapharyngeal space, nor any signs of bony erosion.

revealing an aggressive pilomatrixoma. A limited excision being first performed, we therefore decided to realize a larger skin excision (no residual tumor was found). Six years after the treatment, the patient showed no trace of recurrence.

3. Discussion

The development of the pilomatrixoma could be explained by a disturbance of the hair follicle cycle, in which limited cytological differentiation of pilar keratinocytes occurs (Fig. 5). It is one of the most common

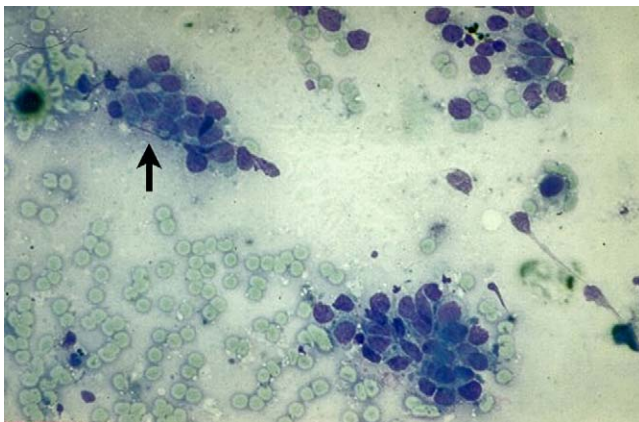


Fig. 2. Fine-needle aspiration biopsy (FNAB): this picture shows various cellular clusters made of cohesive cells (arrow). These also feature a high nuclear to cytoplasm ratio ($\times 400$).



Fig. 3. The lesion has irregular border (arrow) and is made of solid aggregations composed of basaloid cells (arrow) mixed up with cornified material. Note the surrounding fibrous stroma ($\times 100$).

cutaneous appendage tumors in patients aged 20 or less. Pilomatrixoma was first described in 1880 by Malherbe and Chenantais as calcifying epithelioma [3]. Its origin was disputed for decades and in 1949, Lever and Geisner identified hair matrix cells as the origin of these tumors [4]. In 1961, Forbis and Helwig published a histological review of 228 cases and introduced the name pilomatrixoma [5]. In 1966, Reed and Lamar reported on 14 patients with neoplasms that they called “invasive pilomatrixoma” [6]. In 1986, Nield et al. described a particular neoplasm of the preauricular area that showed, not only hair matrical differentiation, but also perineural and vascular involvement. These authors used the term “aggressive pilomatrixoma” for this unusual neoplastic disorder [1]. Pilomatrix carcinoma has rarely been reported and histological criteria for diagnosis are actually not well defined. The present report constitutes, thus, an excellent opportunity to describe a new case of aggressive pilomatrixoma and also to clarify the histological characteristics of those entities.

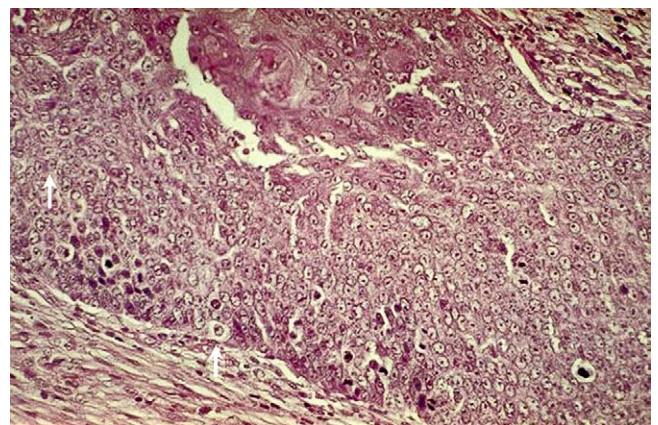


Fig. 4. Basaloid cells demonstrate some cellular atypias and a prominent mitotic activity (arrow) ($\times 400$).

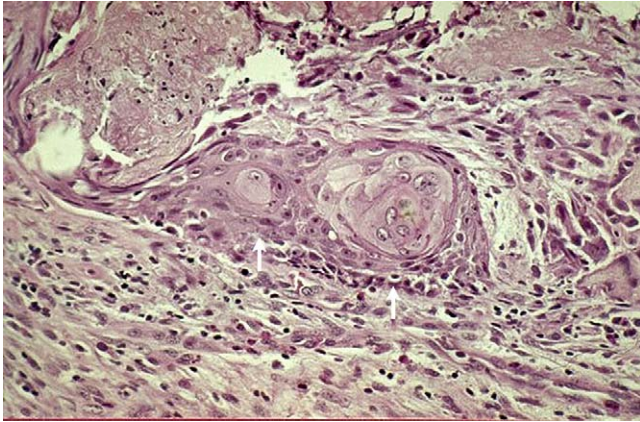


Fig. 5. View of a cellular aggregation demonstrating squamous cell differentiation (arrow) ($\times 400$).

Clinically, pilomatrixomas are slow-growing, benign dermal or subcutaneous tumors, of 0.5–5.0 cm in diameter. These tumors are most frequently found in young patients. Indeed, 40% of cases appear in patients aged 10 or less, and more than 60% are diagnosed during the first two decades of life. The male/female ratio is approximately 2/3. This tumor affects (in decreasing order of frequency) the head, upper extremities, neck, trunk and lower extremities [2,7]. A familial occurrence or an association with myotonic dystrophy has also been reported [8]. Macroscopically, most tumors are rather circumscribed nodules or cystic masses involving the dermis or the subcutaneous tissue. Their consistency varies from soft and friable to firm or hard following a calcifying process.

Histologically, aggressive pilomatrixomas are characterized by atypical and highly proliferating basaloid cells that are disposed in sheets, irregular islands or bands. The latter infiltrate the surrounding tissues. This pattern of infiltration is responsible for local recurrence in case of incomplete surgical excision [9]. The presence of two distinct cell populations, i.e. the anucleated shadow cells and the basaloid cells is fairly specific for pilomatrixoma. Those appear either as isolated cells or in small clusters of two or three. A clear, usually well-defined unstained area occupies the original site of the nucleus, but occasionally a pale residual shadow of the nucleus is still visible. The majority of the basaloid cells are disposed in clusters or in sheets. Basaloid cells are oval to polygonal with poorly defined cell borders and a high nucleocytoplasmic ratio. The “shadow” squamous cells and basaloid cells are surrounded by a background of inflammatory cells and fragments of calcified material. These refractile deposits appear purple to blue, have irregular shapes and sizes but do not show any lamellations suggestive psammoma body formations. Histologically, aggressive pilomatrixoma may be confused with basal cell carcinoma, proliferating pilar cyst and pilomatrix carcinoma [9].

Pilomatrixoma (PMX) is known to be a diagnostic pitfall. Any experienced cyto-pathologist performing FNAs of

superficial, cutaneous lesions is aware of the challenge in differentiating PMX with other benign skin lesions or in falsely diagnosing PMX as a carcinoma. As to underline the reality of this problem of diagnosis, several reports on the cytological features of PMX have been published where up to 45% of cases of PMX were incorrectly diagnosed as other benign tumors (e.g., cysts, adnexal tumors, granulomatous inflammation) or as malignant tumors (e.g., squamous cell carcinoma, small round blue cell tumors, malignant skin appendage tumors) [10–11]. Besides, such diagnostic mistake based on FNA had also been realized in our case.

The typical cytological findings of PMX include a variety of cellular components like basaloid cells, ghost (or shadow) cells, foreign body-type giant cells, nucleated squamous cells and calcium deposits frequently on a background of amorphous debris and chronic inflammatory cells [10]. Basaloid cells and ghost cells appear to be the two key components, as each of these cell-types needs to be present in order to make the adequate cytological diagnosis. PMX do have a variable proportion of diverse cell types. Differential diagnosis of PMX using FNA includes a wide variety of benign and malignant lesions. When the smears show a predominance of ghost cells, differential diagnosis with squamous-lined cysts, such as epidermal inclusion cyst (EIC), trichilemmal cyst (TC) and branchial cleft cyst (BBC) should be considered [10]. A ruptured EIC is more likely to be confused with PMX due to the presence of a foreign body granulomatous reaction with multinucleated giant cells, but basaloid cells and calcific debris are rarely identified in EICs. In TC, in addition to anucleated squamous, FNAs may yield groups of maturing basaloid cells which show lower nuclear to cytoplasmic ratios, denser cytoplasm, better-defined cell borders and inconspicuous to absent nucleoli. Calcific debris and keratin clumps can be quite extensive and when rupture of the cyst occurs, foreign body giant cell reaction can be observed. Concerning malignant lesions, PMX must be distinguished notably from squamous cell carcinoma, Merkel cell tumor (MCT) and small round blue tumor. PMX is occasionally misinterpreted as squamous cell carcinoma, but the young age of most patients, uniformity of the basaloid cells and the lack of significant nuclear atypia in the nucleated squamous cells facilitate differentiation between these two entities. Like PMX, the head and neck area is a common location of Merkel cell tumor, a rare small-cell neuroendocrine carcinoma of the skin. However, MCT occurs in more elderly patients and is locally aggressive. In contrast to PMX showing a polymorphous cell population, FNA smears of MCT demonstrate a monomorphic population of dispersed atypical small blue cells with coarsely granular, irregularly distributed chromatin and inconspicuous nucleoli. Moreover, shadow cells, nucleated squamous cells and multinucleated giant cells are not identified in MCT. In child, small round blue cell tumors must be ruled out when the basaloid cells predominantly occupy the FNA smear [10]. Finally, the differential diagnosis of PMX also includes inflammatory and infectious lesions (reactive

Table 1

Histologic features	Pilomatrixoma	Proliferating/ Aggressive pilomatrixoma	Pilomatrix carcinoma
Basaloid cells	++	++	++
Shadow cells	++	++	++
Nucleo/cytoplasmic Ratio	low	high	high
Capsular effraction	no	yes	yes
Perineural and vascular involvement	no	No	yes
Local invasion	no	yes	yes
Distant invasion	no	no	yes
Central necrosis	no	yes	yes

lymph node or granulomatous lymphadenitis) when the FNA smear shows abundant chronic inflammatory cell infiltrate with lymphocytes, histiocytes and macrophages.

So, accurate preoperative diagnosis of PMX by FNA cytology can be made with confidence if a combination of basaloid cells, ghost cell, nucleated squamous cells and foreign-type giant cells are encountered in FNA smears from a skin-based nodule. Moreover, basaloid cells and ghost cells appear to be the key components as the presence of these two cell types without nuclear atypia is fairly specific.

Finally, the limit between aggressive pilomatrixoma and pilomatrix carcinoma is still not clear. According to Inglefield et al., the aggressive pilomatrixoma key histological features are a high mitotic rate and the presence of excessive basaloid cell proliferation with discrete nodules distant from the main lesion [9]. In this regard, Kaddu et al. have described a distinctive proliferative variant of pilomatrixoma and proposed the designation “proliferating pilomatricoma” [12]. Our point of view concerning proliferating and aggressive pilomatrixomas is that those entities are morphologically very close and characterized by a local invasion and their capacity to recur [9,12]. The Table 1 constitutes an attempt to clarify the limits between those entities. Pilomatrix carcinoma is formed by irregular

shaped cellular bands and sheets of basaloid cells presenting hyperchromatic nuclei and prominent nucleoli. In the central part, necrosis, keratin and shadow cells are often seen. Both immunohistochemical and flow cytometric analyses have been performed to establish whether these methods may be used to differentiate pilomatrix carcinomas from its benign counterpart. But, neither of these methods has been successful. The main indicators of malignancy appear to be nuclear pleomorphism, frequent and atypical mitosis, central necrosis, infiltration of skin, soft tissue, blood and lymphatic vessels and ulceration [13]. Pilomatrix carcinomas are locally aggressive tumors, tending to recur when incompletely excised. Rarely, metastasis and death can result from these tumors.

Wide surgical excision is the best treatment against aggressive pilomatrixoma and pilomatrix carcinoma. The efficiency of radiation therapy is unclear as the experience in this type of treatment is still very limited. Careful histological study of resection margins as well as patient follow-up is indicated as recurrences are frequent but subject to therapy [13].

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