

Pilomatricoma of the Auricular Region: Case Report

Tarcília Aparecida da SILVA¹
Edgard Franco MORAES JÚNIOR²
Alberto CONSOLARO¹
Vanessa Soares LARA¹

¹Department of Stomatology (Pathology), Faculty of Dentistry of Bauru, University of São Paulo, Bauru, SP, Brazil

²Discipline of Bucco-Maxillo-Facial Surgery and Traumatology and Implantodontology, FISA/FUNEC, Santa Fé do Sul, SP, Brazil

Pilomatricomas are relatively rare tumors of ectodermal origin from the outer root sheath cell of the hair follicle. They are usually asymptomatic, solitary, firm or hard, freely mobile, dermal or subcutaneous nodules. The purpose of this article is to present a case that illustrates the diagnostic difficulty encountered by oral surgeons and pathologists and to review the literature regarding pilomatricomas of the auricular region.

Key Words: pilomatricoma, hair follicle tumors.

INTRODUCTION

Pilomatricoma usually presents as an asymptomatic, solitary, firm or hard, freely mobile, dermal or subcutaneous nodule. The tumor generally exhibits no fixation to neighboring tissues and has an osseous- or cartilage-like hardness. The overlying skin surface is typically flesh colored, but may be erythematous or bluish. Unusual clinical variations include large extruding or perforating examples, multiple eruptive cases, familial cases and malignant examples (1,2).

There is a predilection for the head, neck and upper extremities with approximately half of all reported cases occurring in the head and neck (1-4). Some cases have been closely associated with structures of the oral cavity. The most frequently reported site of pilomatricomas of the oral region is the auricular region with a frequency of 33-79% (3,4). In some cases, these lesions may simulate a neoplasm of the parotid (5-16).

A search of the English literature revealed 10 well-documented cases of pilomatricomas in the auricular region; these as well as the current case are presented in Table 1. Additionally, 71 cases of pilomatricomas in the auricular region have been reported by Makek et al. (10) (20 cases), Rink (3) (6

cases), Yoshimura et al. (4) (22 cases) and Danielson-Cohen et al. (15) (23 cases), although details of these cases were not provided.

CASE REPORT

A 25-year-old white woman was referred for evaluation of a swelling in the right auricular area. The patient had noted the auricular mass 15 years before initial presentation. The lesion had begun growing over the last few months. There was no history of pain, infection or trauma, and no history of facial nerve dysfunction. Clinical examination revealed a solitary, firm, well-circumscribed, freely mobile mass located below the right ear. The overlying skin was normal in appearance. The mass was fixed to the underlying tissue, and bimanual palpation failed to localize the origin of tumor. The parotid duct was unremarkable and salivation was normal. A preoperative radiograph revealed a sharply outlined subcutaneous mass, which was calcified in its central portion (Figure 1). Fine-needle aspiration demonstrated scattered erythrocytes and lymphocytes.

A preoperative diagnosis of benign tumor was proposed. The surgical procedure consisted of enucle-

ation of the lesion using an extraoral approach. At the time of surgery, the tumor was easily shelled out of the surrounding tissues (Figure 2). The surgical specimen revealed a hard irregular mass measuring 1.5 x 1.0 x 0.8 cm, and was yellowish-white in color (Figure 3). Microscopically, the lesion was well circumscribed and composed of irregularly shaped masses of shadow or ghost cells, scattered basophilic cells that undergo abrupt keratinization forming shadow cells and areas of ossification. The basophilic cells were oval to polygonal with poorly defined cell borders, a scant cytoplasm, intensely staining basophilic nuclei and high nuclear-cytoplasm ratio. Shadow cells showed dark granularity and lack of staining in the area previously occupied by their nuclei. Sparse multinucleated giant cells were observed; however, no considerable foreign-body reac-

tion was seen in stroma (Figure 4). Follow-up at 2 years revealed no evidence of recurrence.



Figure 1. Panoramic radiograph shows a circumscribed radiopaque structure at the right auricular region (arrows).



Figure 2. Transoperative view of the tumor in the right auricular area.



Figure 3. Surgical specimen with multilobulated and capsular appearance measuring 1.5 x 1.0 x 0.8 cm

Table 1. Cases of pilomatricomas in the auricular region reported in the literature.

Reference	Gender	Age	Ethnic group	Duration	Size	Microscopic findings	Preoperative diagnosis
Gullane et al. (8)	F	12	White	6 m	1 cm	SC, MGC, K	NR
Brandner and Bunkis (5)	M	8	Hispanic	8 m	6 x 5 x 4 cm	BA, K	Parotid neoplasm
Makek et al. (10)	F	14	White	4 m	2 x 1.5 cm	B, MGC, SC	Carcinoma
Yoshimura and Oka (14)	F	14	Japanese	2 y	3 x 2.5 x 1.3 cm	BA, SC	Benign calcified tumor
Ooil et al. (11)	M	19	Chinese	3 m	6 cm	B, BA, MGC	Skin tumor; hemangioma
Thomas et al. (12)	M	16	NR	18 m	2 cm	NR	NR
	M	19	NR	NR	NR	NR	Pilomatricoma
	F	25	NR	NR	NR	NR	NR
Phyu and Bradley (16)	M	51	White	14 m	1.5 cm	NR	Pilomatricoma
Present case	F	25	White	15 y	1.5 x 1.0 x 0.8 cm	SC, O	Benign tumor

F, female; M, male; NR, not reported; B, basaloid cells; BA, basophilic cells; K, keratinization; MGC, multinucleated giant cells; O, ossification; SC, shadow cells. No symptoms were reported for any of the cases.

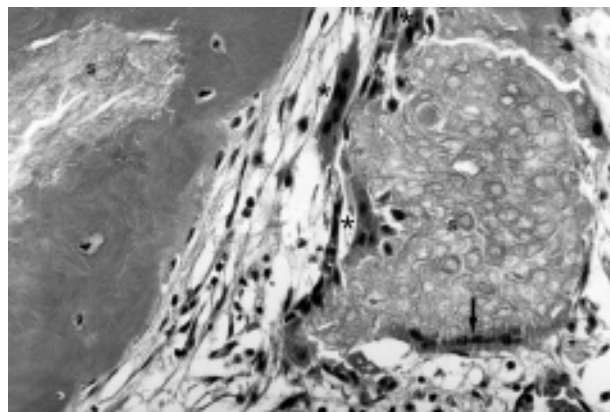


Figure 4. Photomicrograph showing devitalized island of ghost or shadow cells (S) involved by a fibrous stroma. Peripherally, the transition between basophilic and shadow cells (arrow) is observed. Sparse multinucleated giant cells are seen (*). On the left, a group of shadow cells is seen adjacent to a foci of ossification. (Hematoxylin and eosin; original magnification: 100X).

DISCUSSION

Pilomatricomas are of ectodermal origin and arise from the outer root sheath cell of the hair follicle (1). The microscopic appearance of pilomatricomas is characterized by irregularly shaped well-circumscribed masses of epithelial cells. Basophilic cells and shadow cells may be observed in the cellular stroma. In some tumors, the basophilic cells are rare as seen in the present case. It is often possible to observe the transition of basophilic cells to shadow cells (1,17,18). As proposed by Fayyazi et al. (17), immature basophilic cells differentiate after a temporary cell cycle arrest into either squamoid cells or transitional cells. Whereas the squamoid pathway represents complete epithelial differentiation, the transitional pathway represents a deteriorate trichocytic differentiation leading to apoptosis and formation of dead shadow cells. Areas of keratinization, calcification and ossification may be seen in the stroma. In the present case, only areas of ossification were observed (Figure 4). The stroma often contains a chronic inflammatory cell infiltrate with foreign body giant cells. Mitotic figures may be numerous. Active proliferating hyperchromatic cells with numerous mitoses and infiltration of surrounding structures are microscopic features suggestive of malignant pilomatricomas (1,17,18).

Four distinct morphological stages of pilomatri-

comas have been proposed (18): a) early: small and cystic lesions; b) fully developed: large and cystic neoplasms; c) early regressive: foci of basaloid cells, shadow cells and lymphocytic infiltrate with multinucleated giant cells; d) late regressive: numerous shadow cells, absence of basaloid or inflammatory cells; calcification and ossification may be present. The natural history of the neoplasm is thought to be a developmental process in which the lesion begins as an infundibular matrix cyst and ends up as a calcified or ossified nodule. By using the aforementioned criteria, the present case appeared to be in the late regressive stage, which is consistent with the clinical history of being present for 15 years.

Making a clinical diagnosis of pilomatricoma can be difficult, although the presence of a nodule of this type, especially in younger patients, should raise suspicions of pilomatricoma. The differential diagnosis of these lesions should include sebaceous, dermoid and epidermoid cysts, metaplastic bone formation, foreign body reaction, parotid gland tumor, hematoma, osteochondroma, trichoepithelioma and basal cell epithelioma (9,10,13). Most pilomatricomas do not cause clinical complications. Due to slow growth, a considerable time usually elapses before diagnosis resulting frequently in large tumors. The clinical course is generally benign although, malignant transformations have been reported (1,2). In some instances, the original diagnosis of pilomatricoma in those cases with "malignant transformation" might be questionable.

Recommended management is surgical excision. Recurrence is uncommon after adequate excision. All patients need dermatological evaluation with close long-term follow-up. Analysis of cases (Table 1) shows that pilomatricoma was included in the preoperative diagnosis of only two cases (12,16). Oral surgeons should expand their differential diagnosis of calcified masses in the auricular region to include pilomatricomas.

RESUMO

Pilomatricomas são tumores relativamente raros de origem ectodérmica a partir das células da bainha externa do folículo piloso. Estes se apresentam geralmente como nódulos dérmicos ou subcutâneos, usualmente assintomáticos, solitários, firmes ou duros e extremamente móveis. O objetivo deste artigo é apresentar um caso que ilustra a dificuldade de diagnóstico encontrada pelos cirurgiões e patologistas bucais e apresentar uma revisão da literatura relativa aos pilomatricomas da região auricular.

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