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Case Report

Case Report and Highlight Clues on the Diagnosis of Pilomatrical Carcinoma

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ABSTRACT

Pilomatrical Carcinoma (PC) is a rare malignant adnexal tumor with matrical differentiation. Its benign counterpart (Pilomatrixoma) is diagnosed much more frequently in daily pathological practice. Both entities share genetic alterations but the malignant counterpart acquires mutations that makes it develop an aggressive behavior [1].

We describe a 33-year-old man who presented with a 7 x 6 cm nodular ulcerated lesion in the left ear with markedly accelerated growth in the last month. Incisional biopsy was referred to us with suspicion of squamous cell carcinoma versus pyogenic granuloma. Histologic sections showed ulcerated fragments infiltrated by a basaloid cell proliferation interspersed with groups of "ghost cells". The neoplastic cells were arranged in irregular sheets with infiltrative borders. Groups of Squamous cells with trichilemmal keratinization and foci of necrosis were also identified. The biopsy was diagnosed as an adnexal neoplasm with pilomatricial differentiation, suggesting its complete resection with safety margins due to the presence of aggressive characteristics. The subsequent study of the excisional biopsy showed similar characteristics to those previously described. Notoriously, focal infiltration of the auricular cartilage was identificated, leading us to the undoubted diagnosis of pilomatrical carcinoma.

Keywords: Pilomatrical Carcinoma; Adnexal Cutaneous Neoplasm; Pilomatrixoma

Introduction

Pilomatrical carcinoma (PC) is a malignant adnexal neoplasm with matrical differentiation that shows a very low incidence. There are about 150 cases reported in the literature so far. There are no risk factors for these tumors, around 60% of these cases present in the head and neck region [2].

Clinically and epidemiologically, in contrast to pilomatrixoma, PC usually occurs more frequently in men during the fifth to seventh decade of life, as a large tumor (up to 10 cm) with poorly defined margins and with an ulcerated surface. According to the literature, both entities are related and share similar genetic alterations, but PC probably acquires additional mutations over time that endow it with a more aggressive behavior [1].

Limited biopsy may miss key histopathological features because of limited tissue sampling and make the diagnosis considerably challenging [3].

Because of the malignant nature of the lesion, surgical treatment it is usually performed. Adequate margin resection measurement is controversial. Margins between 5mm-2cm are reported as sufficient. In some cases, Mohs surgery has been described as an alternative approach [2,4].

As informed in the literature, 23% of patients may develop local recurrence within the first 7 months despite the presence of clear surgical margins [2].

Metastatic disease is extremely rare, being lymph nodes and lungs the most frequent location [1,5].

Case Presentation

We describe a 33-year-old male who was admitted to our hospital with non-previous medical condition, consulting for a tumoral lesion noticed since an indeterminate time ago. It remained stable and invariable for several months but unexpectedly an accelerated an exponential growth occurred during the last month.

On physical examination, a 7 x 6 cm brownish ulcerated firmly attached nodular tumor was recognized. It compromised almost the entire surface of the left outer ear and showed poorly defined edges (Figure 1). An incisional biopsy was referred to our service for histopathological study with clinical suspicion of squamous cell carcinoma versus pyogenic granuloma.

The microscopic study revealed an ulcerated fragment covered by fibrinoid material and abundant granulation tissue (Figure 2) infiltrated by a proliferation of basaloid cells with scant cytoplasm and hyperchromatic, round to oval nuclei (Figure 3). The cells were arranged in irregular sheets, nests and broad cords with an infiltrative growth pattern (Figure 4). They alternated with groups of “ghost cells” with extensive eosinophilic cytoplasm, surrounded by foreign body reaction with giant cells (Figure 5), and squamous cells with trichilemmal keratinization (Figure 6). Additionally, osseous metaplasia (Figure 7) and foci of necrosis and suppuration were identified (Figure 8).

The lesion was diagnosed as an adnexal neoplasm with pilomatrical differentiation, suggesting a complete resection with safety margins due to the presence of aggressive characteristics such as necrosis, infiltrative border, size and ulceration.

The excisional specimen study showed similar characteristics to those previously described and in addition we could certify focal infiltration of the cartilage (Figures 9 and 10). These findings reaffirmed the diagnosis of PC.

Conclusion

PC is a rare entity with few cases reported in the literature. It is considered a low-grade neoplasm, with substantial rates of local recurrence but low frequency of metastases, mainly to lymph nodes and lungs [5].

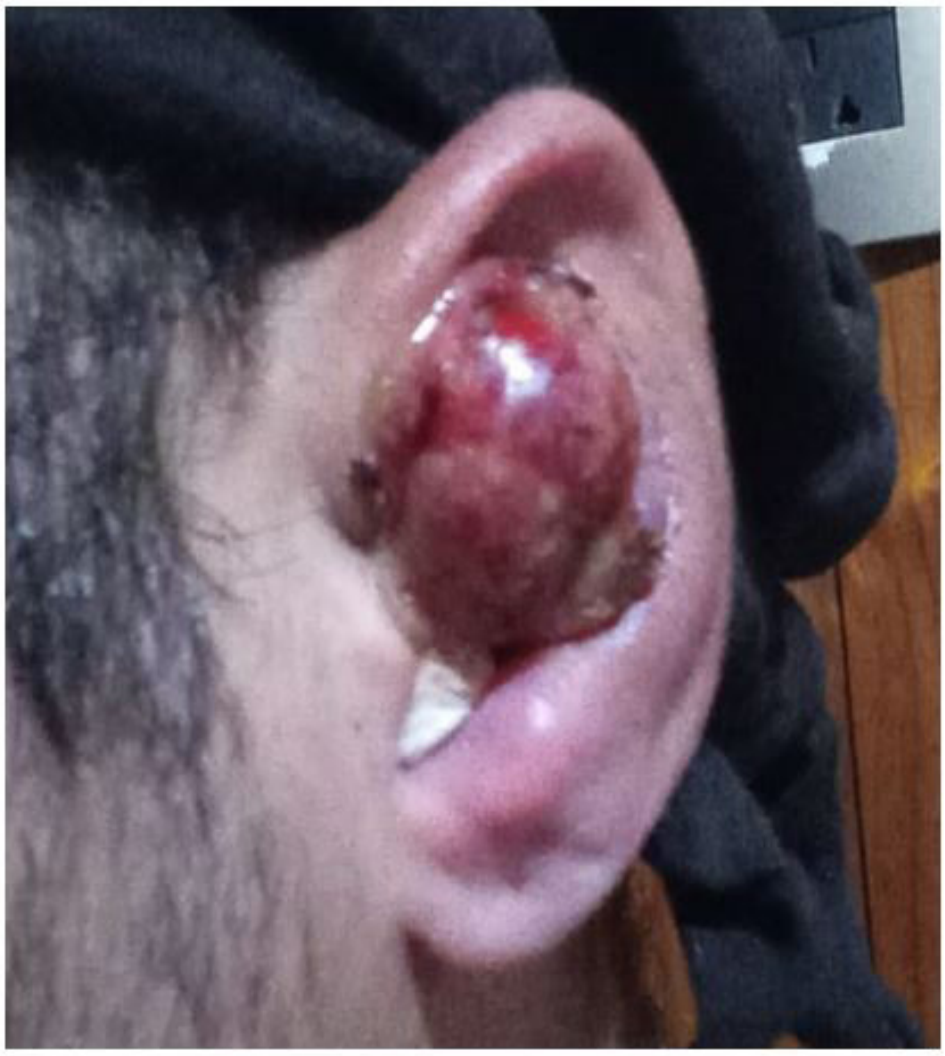

Figure 1: Clinical image of the ulcerated nodular tumor in the outer left ear

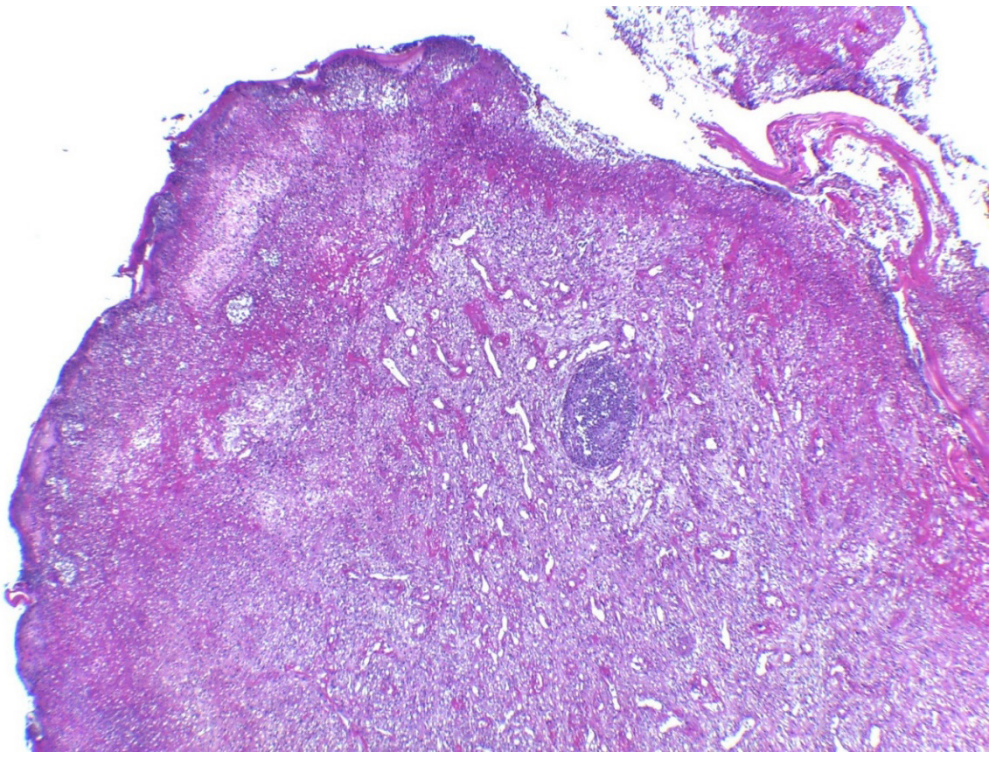

Figure 2: H&E 40x: Ulcerated fragment covered by fibrinoid material and abundant granulation tissue

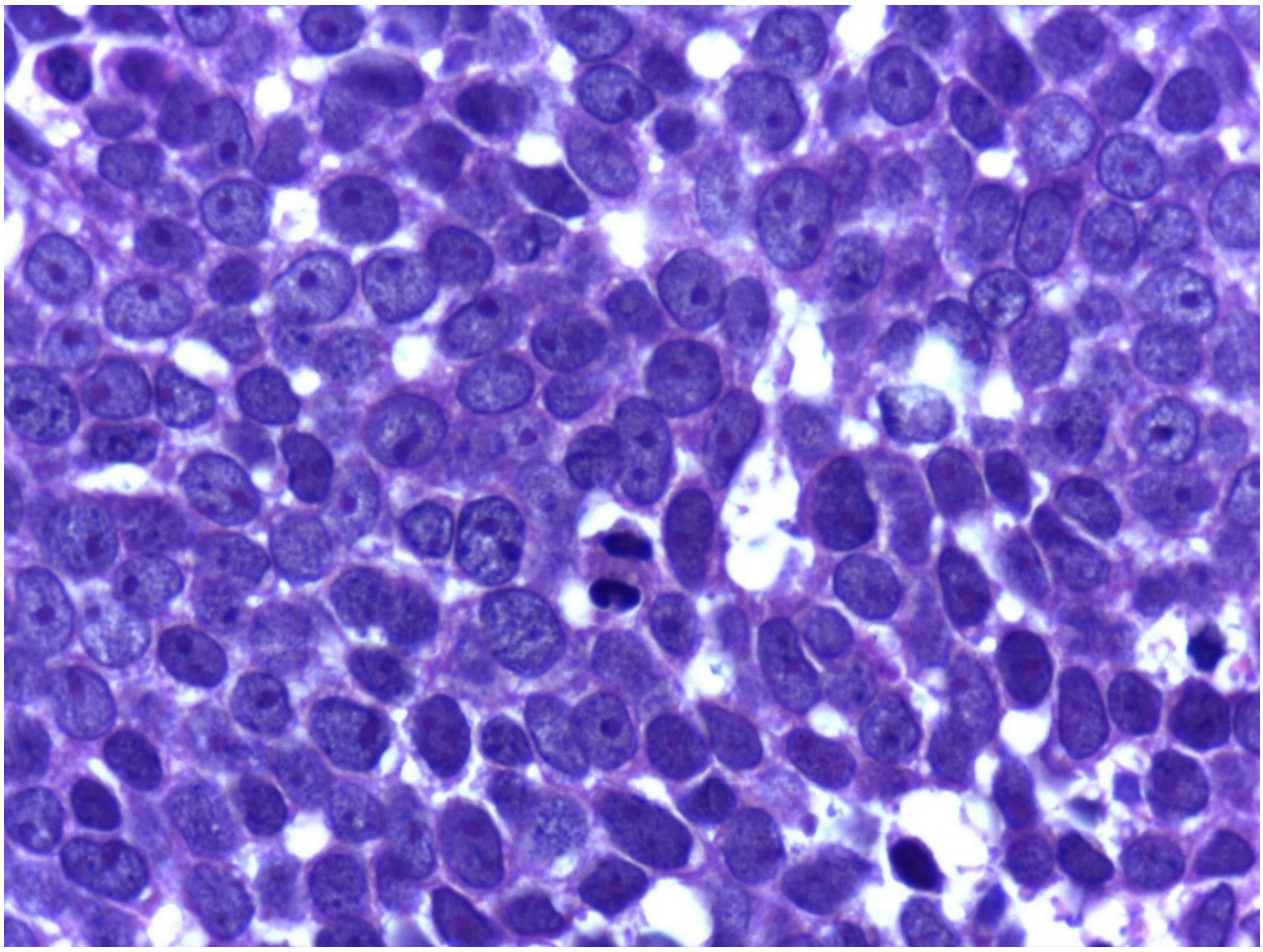

Figure 3: H&E 1000x oil: Proliferation of basaloid cells with scant cytoplasm and hyperchromatic, round or oval nuclei

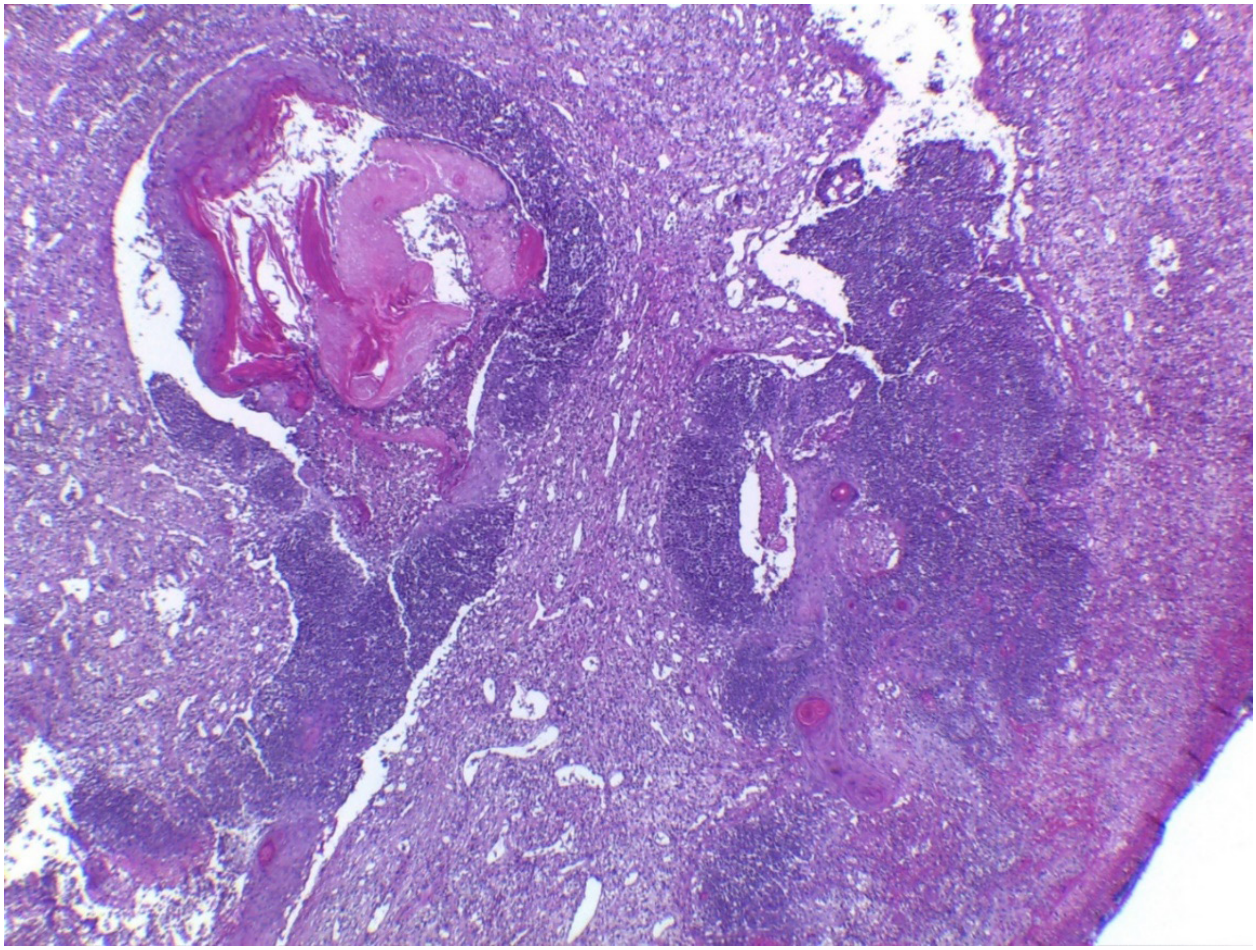

Figure 4: H&E 40x: Proliferation arranged in nests and broad cords, with infiltrative-like growth margins

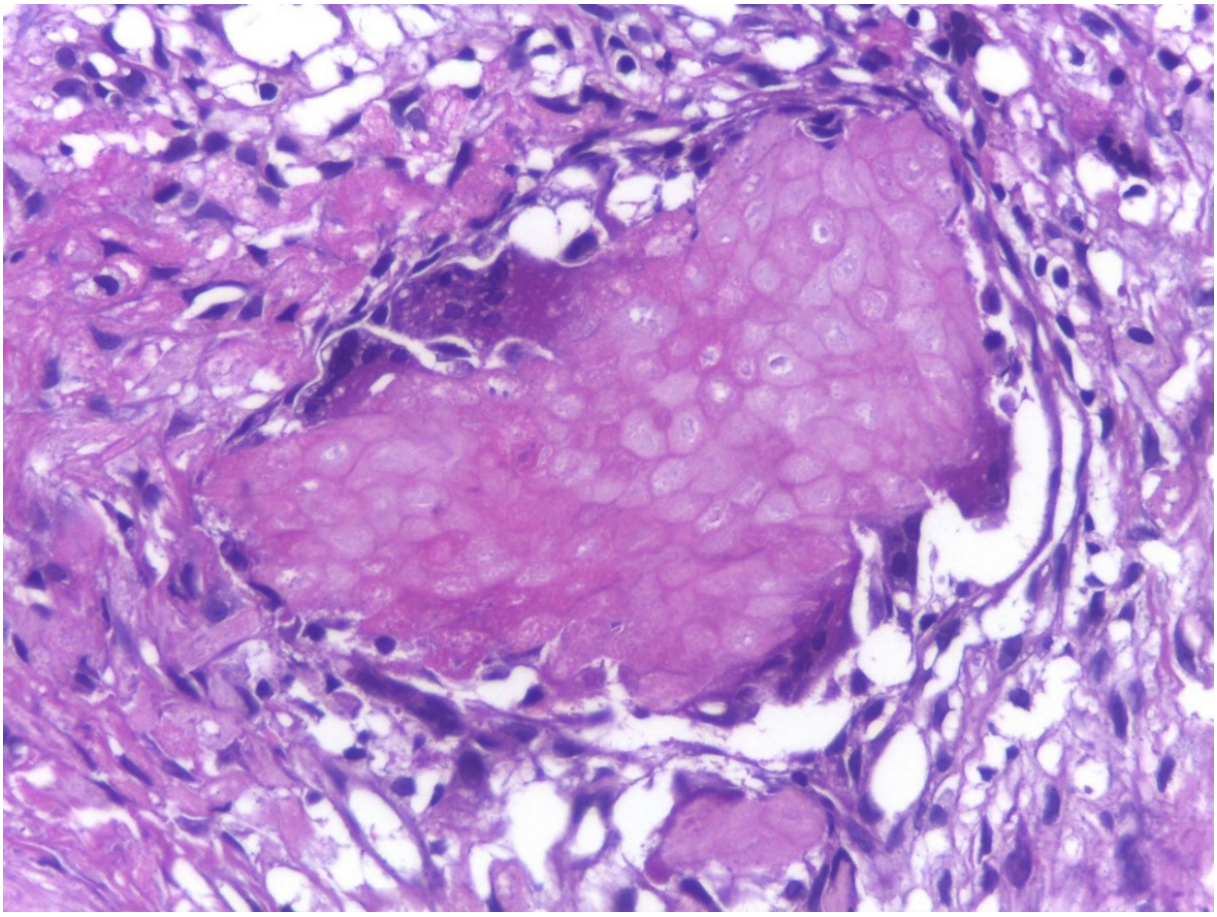

Figure 5: H&E 100x: Groups of “ghost cells” with extensive eosinophilic cytoplasm, surrounded by a foreign body reaction

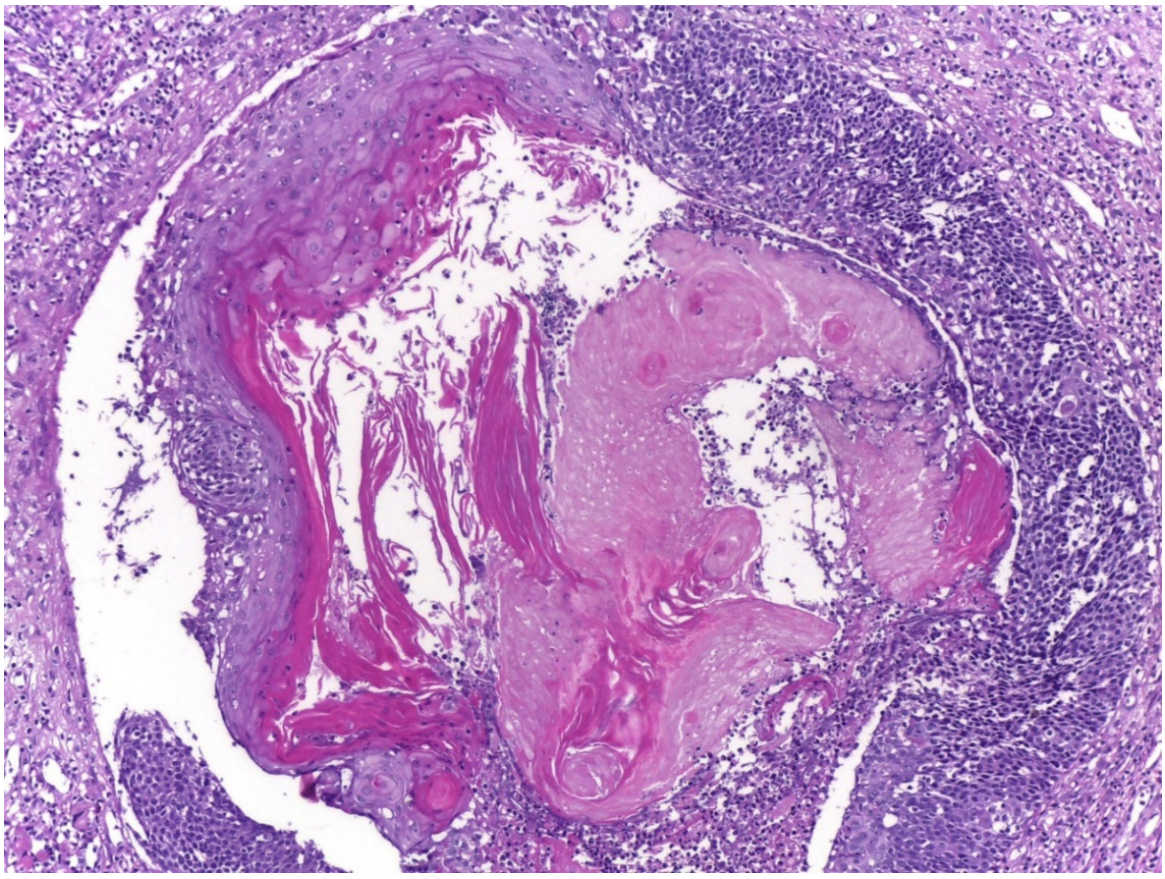

Figure 6: H&E 100x: Squamous cells with trichilemmal keratinization

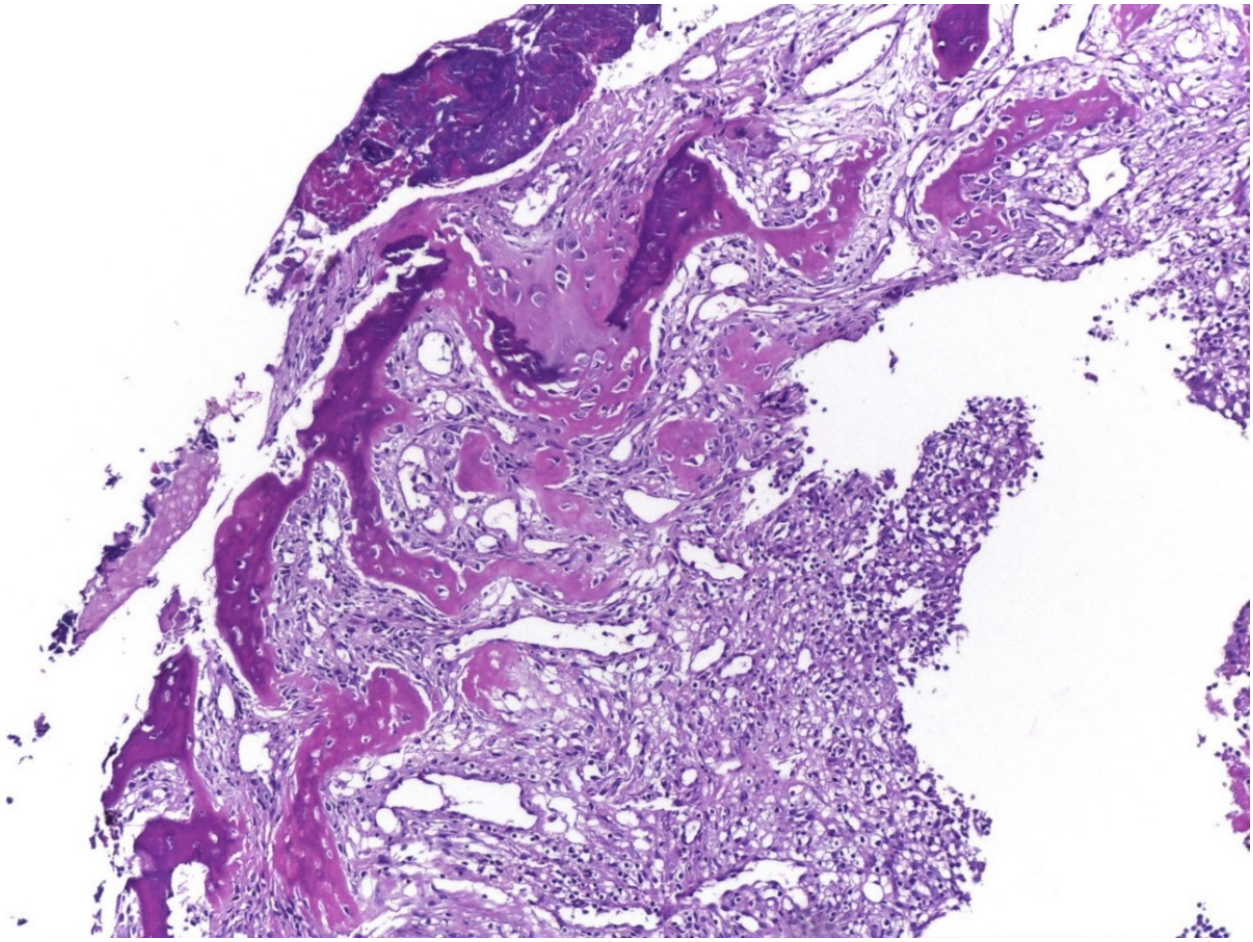

Figure 7: H&E 40x: Osseous metaplasia

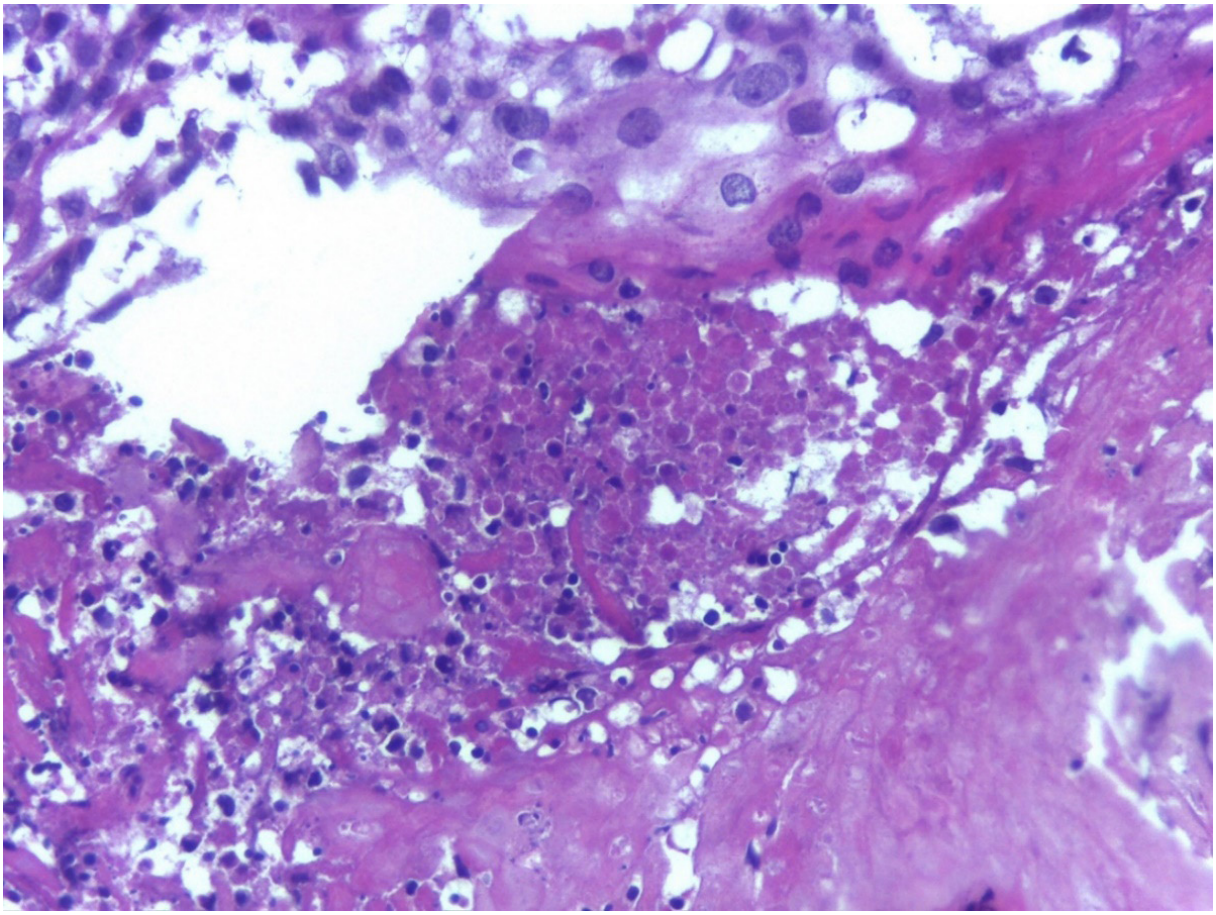

Figure 8: H&E 400x: Foci of necrosis

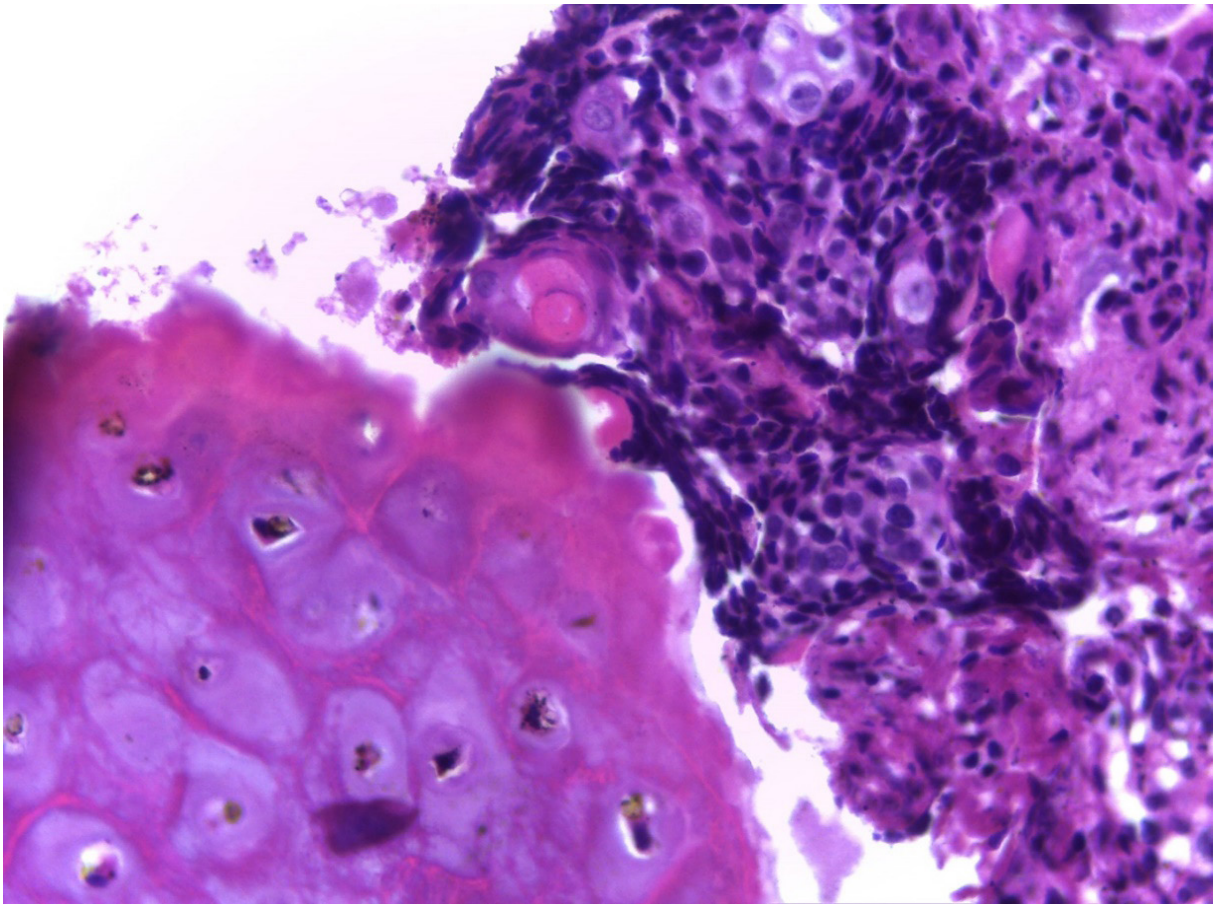

Figure 9: H&E 40x: Basaloid neoplasm infiltrating cartilague

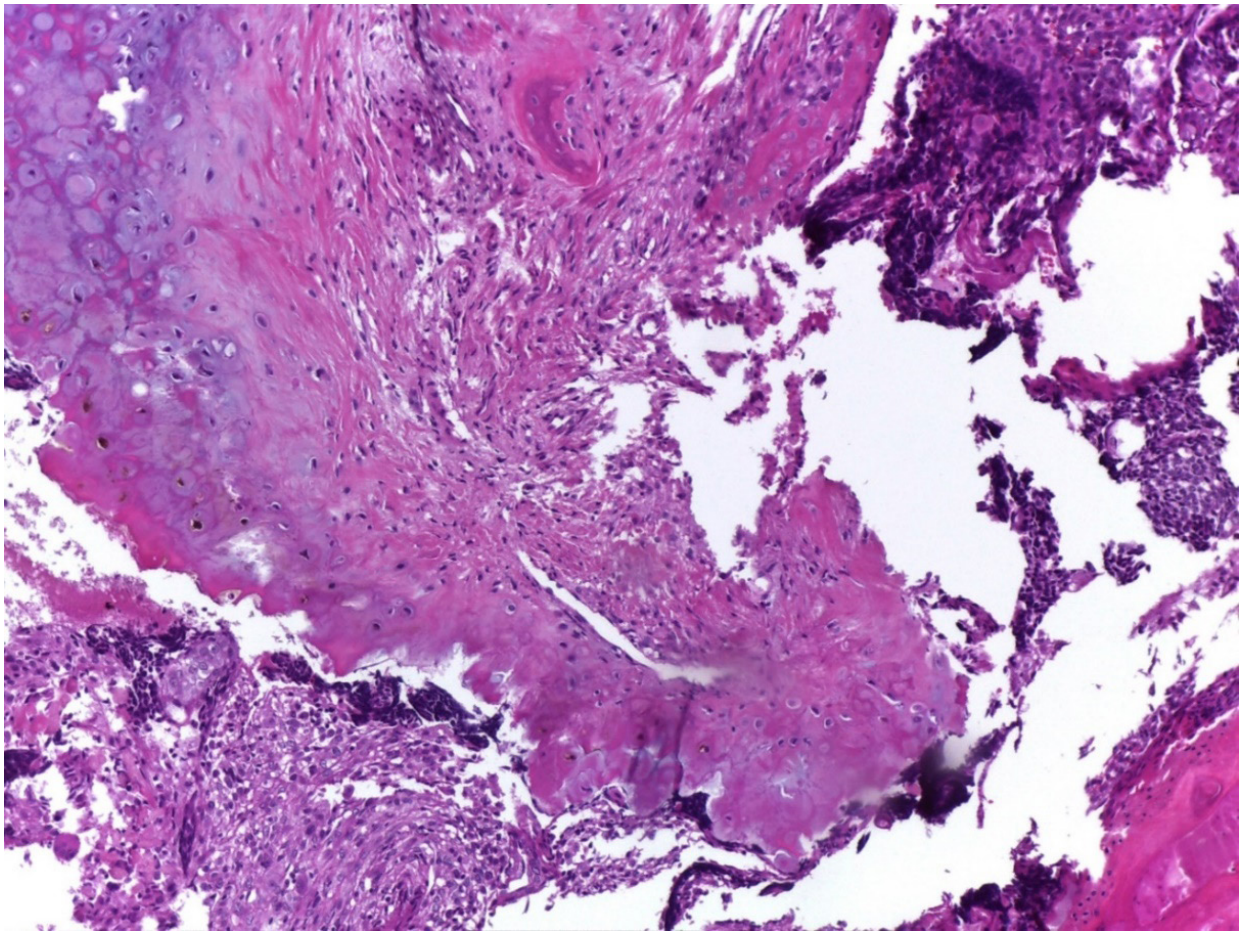

Figure 10: H&E 1000x oil: Basaloid neoplasm infiltrating cartilague

Clinical and histological features are diverse and simulate other tumours (pilomatrixoma, basal cell carcinoma with matrical differentiation, etc.), that can lead to diagnostic errors. It is appropriate to seek the clinical and pathological characteristics of aggressiveness. Macroscopic and microscopic features suggestive of malignancy include size greater than 4 cm, necrosis, infiltrative borders, predominance of basaloid cells, atypical mitoses, nuclear atypia, compromise of deep tissue, vasculolymphatic and perineural invasion [6]. The immunohistochemical profile has been studied to differentiate benign and malignant counterparts but was not useful to make this distinction.

It is important to highlight certain situations in particular, as happened in our case, were in first instance we only received a small and incisional biopsy. In such cases it might not be possible to find all malignancy characteristics but at the slightest suspicion of aggressive behavior it is relevant to report it and suggest a wider resection.

We presented a novel case of Pilomatricial Carcinoma with a suggestive but not conclusive diagnosis in the initial biopsy and we also include wise advices when facing this situations to achieve the best medical practice, treatment and patient follow-up.

References

1. Jones C, Twoon M, Ho W, Portelli M, Robertson BF, et al. (2017) Pilomatrix carcinoma: 12-year experience and review of the literature. J Cutan Pathol 45: 33-8.

2. Gaurav M, O'connor J, Pappa E, Pollock J (2020) A Rare Neglected Pilomatrix Carcinoma on the Forehead: A Case Report. Case Reports in Dermatology 5: 10.35248/2684-124X.2020.5.163.

3. Lohiya S, Rawal YB, Dillon JK (2018) Giant Pilomatrix Carcinoma of the Face. J Oral Maxillofac Surg 76: 368-73.

4. Papadakis M, de Bree E, Floros N, Giannikaki E, Xekalou A, et al. (2017) Pilomatrix carcinoma: More malignant biological behavior than was considered in the past. Mol Clin Oncol 6: 415-8.

5. Eduardo Calonje J, Brenn T, Lazar AJ, Billings S (2012) McKee's Pathology of the Skin (4th Edn) Volume 2, Elsevier, USA.

6. Herrmann JL, Allan A, Trapp KM, Morgan MB (2014) Pilomatrix carcinoma: 13 new cases and review of the literature with emphasis on predictors of metastasis. J Am Acad Dermatol 71: 38-43.e2.