Pilomatrix Carcinoma: A Case Report

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Abstract

Pilomatrix carcinoma is a rare malignant tumor. It is usually reported in patients over 50 years. Nowadays, there is no standardized treatment while a high risk of recurrence after simple surgery was frequently observed. We report one case of a pilomatrix carcinoma in a 15 years old patient. In spite of a surgery with wide margins, she presented a local recurrence on the head and a lung metastasis. She had undergone a second surgery with additional radiotherapy and chemotherapy. She is now in remission.

Introduction

Pilomatrix carcinoma was described in 1880 by Malherbe, is a rare and malignant adnexal tumor. It is a dermo-hypodermic tumor, with a low metastatic potential but a high risk of recurrence after excision. The average age of occurrence is 46.3 years [1]. We report a case of a pilomatrix carcinoma of the head in a 15 years old patient.

Case Report

A 15-year-old woman presented a 2cm lesion of the head, which was surgically removed in a local clinic. The lesion was sent to a dermatopathologist who diagnosed a pilomatrixoma with no evidence of malignancy. Unfortunately, the patient was lost to follow-up. Five months later, she was examined in our hospital for a recurrent lesion, which was burgeoning necrotic and measured 6cm in diameter at the previous scar. The CT scan before surgery showed a hypervascular tumor leading to an angiomatous lesion (Figure 1). The tumor was removed 48 hours after an embolization (Figure 2). Malignant cells were found in the extemporaneous histological examination but an immediate reconstruction with skin graft was practiced at the resection area. Few days later, pilomatrix carcinoma was identified in finally histopathological analysis.

A thoraco-cervical-abdominal CT scan with contrast did not show suspicious lymph nodes or distant metastases. The first surgical samples were reanalysed and read by our pathologists who have found the same characteristics of a pilomatrix carcinoma tumor. The patient was re-operated to get a 2cm margin of healthy tissue around the lesion according to the recommendations of cutaneous oncologists of the Saint Louis hospital (Paris). The peristeme wasn’t invaded. A scalp rotation flap reconstruction has been done immediately to cover the resection area. The patient presented a recurrence 3 months later with an invasion of bone and superior sagittal sinus, and she underwent a third surgery with craniotomy (Figure 3). The CT scan showed a lung...
metastasis. She was treated by chemotherapy with 5-Fluorouracil and cisplatin, then a radiochemotherapy with cisplatin and 60 gray during 30 sessions. After this treatment, the metastasis disappeared and sagittal sinus is permeable. The general condition of the patient is good and the clinical examination is normal. She will undergo a clinical examination by oncologist pediatrician, a cerebral MRI and a thoracic CT scan every two months over the next two years. There is no local recurrent lesion at 6 months after treatment.

Discussion

Malherbe was described Pilomatrix carcinoma for the first time in 1880 as a “calcifying epithelioma” derived from cells of the sebaceous glands [2] but the malignancy has been known only since 1927 [3]. There are about 80 cases reported to date, including ten cases with visceral metastases. The diagnosis of pilomatrixoma is based on histological examination. This is a nodular lesion with basaloid cell component and mummified plant on the outskirts, and eosinophil content. It is accompanied by a fibrous stroma and granulomatous reaction. In order to conclude the malignancy of the lesion, the tumor infiltration associated to the nuclear atypia, the abnormal mitosis and the numerous tumor necrosis must be found in the histological examination [4-7] (Figure 4-7). No risk factor is known to date [6], but these lesions were observed frequently in the white male and over 50% of cases have been reported in patients over 50 years old [2]. More than 60% of cases were on the face and neck [8]. Lung metastases are the most described and they could be diagnosed until four years after the onset of initial injury. Local staging consists of a scanner or an MRI of the lesion. General staging should include palpation of lymph nodes and a cervico-thoraco-abdominal scanner to search for lung and liver metastases. A brain and bone scan should be performed only when there are warning signs [9-11]. Treatment consists of a surgical excision with wide margins (2-3 cm) because of high recurrence rate after simple excision (over 50% of cases). Peritumoral margins are not codified [12]. Radiation therapy, which is not the first-line treatment, is sometimes used before surgery, after surgery or palliative purposes (to be analgesic in secondary locations such as spinal) [13]. Chemotherapy has not been proven effective but it can be used in case of metastases [10,11,14,15].

Conclusion

Pilomatrix carcinoma is a rare malignant tumor. The histological diagnosis, based on several factors (necrosis, nuclear atypia, infiltration and abnormal mitosis) is difficult to prove. Treatment is not standardized. Surgical procedure with wide margins to avoid the large recurrence of relapse is recommended when the staging shows no metastasis.

References


