Cranial vault tumor: malignant proliferating trichilemmal tumor of the scalp

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Proliferating trichilemmal tumor (PTT) is a rare cutaneous tumor that mainly occurs in the scalp, eyelids, neck, and face of elderly women. In most cases, it is a unique, large, multi-lobulated, cystic lesion originating from a hair follicle. These tumors are mostly benign and are characterized by trichilemmal keratinization. However, in rare cases, they may be aggressive with a propensity for distant metastasis.

We report the case of a 69-year-old male patient, who presented with as welling on his forehead that had rapidly increased in size over the past year. CT scan and magnetic resonance imaging (MRI) showed a tumor located on the cranial vault, associated with an intracranial extension. Chest X-ray and CT scan found a tumor in the right lung. The patient underwent surgery for the removal of the tumor. The histological examination confirmed the diagnosis of malignant PTT (MPTT).

To the best of our knowledge, only a few cases of MPTT have been published. Furthermore, only one case of MPTT of the scalp with endocranial extension and lung metastasis has been reported to date.

Through this case, we discuss the prognostic factors and the management of this aggressive lesion.

Keywords: metastases; dermatology, neurosurgery, cranial vault tumor

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INTRODUCTION

Proliferating trichilemmal tumors (PTTs) were first described in 1966 by Jones, who referred to them as "proliferating epidermoid cysts" [1]. They are defined as rare cutaneous neoplasms derived from the outer root sheath of the hair follicle [2]. Histologically, these tumors are characterized by trichilemmal keratinization, which corresponds to a sudden transition from the spinous layer to the stratum corneum because of the absence of the granular layer [3]. PTTs are usually benign, but in some rare cases, they can exhibit malignant behavior, invading neighboring tissues and showing anaplasia and necrosis; these cases are referred to as malignant proliferating trichilemmal tumors(MPTTs), which are invasive and metastatic [4]. To the best of our knowledge, only a few cases of MPTT have already been published [3]. Furthermore, only one case of MPTT of the scalp with endocranial extension and lung metastasis has been reported to date.

Given the very limited number of cases, there is no consensus for the management of MPTTs. We report a case of MPTT with endocranial extension and lung metastasis; we then go on to discuss therapeutic issues and prognostic factors.

CASE REPORT

We report the case of a 69-year-old male long-

term smoker, who presented with a swelling of the scalp that had rapidly increased in size over the previous year. He also complained of headaches for the previous two months, as well as anorexia. No history of irradiation or pre-existing skin lesions was found. Clinical examination revealed a subcutaneous, soft, non-painful mass of the forehead, measuring 22 cm in diameter and fixed to the bone. The skin was thin, shiny, and tense. No inflammation or ulcerations could be seen (Figure 1). This lesion was unique. No locoregional lymphoid nodes were found. The neurological and somatic examinations were otherwise normal.

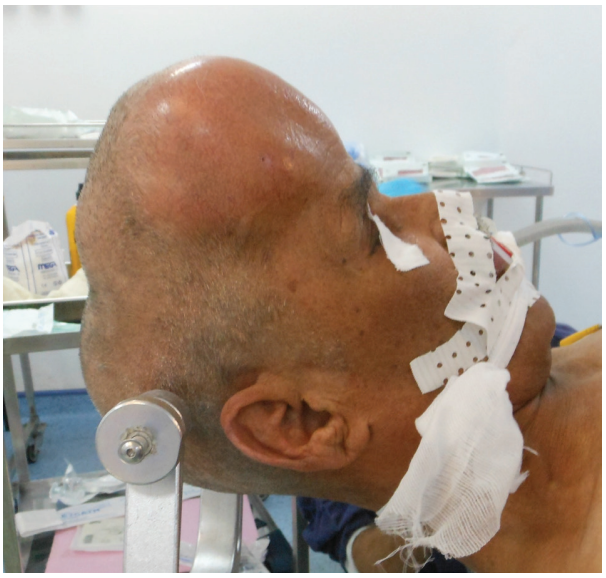

Figure 1. A preoperative photograph: frontal swelling covered with normal scalp measuring 14 cm in axis.

A cerebral CT scan (Figure 2) showed a cystic trabeculated mass, with osteolysis of the underlying vault and dural invasion. Cerebral magnetic resonance imaging (MRI) showed a frontal multi-lobulated cystic mass with a liquid-liquid level. This lesion had invaded the frontal bone and the dura mater, coming into contact with the superior sagittal sinus (SSS), which remained permeable. An enhancement of the cyst wall, septa, and meninges was noticed after the injection of gadolinium (Figure 3). Diagnoses of hydatid cyst of the vault and cystic carcinoma were evoked.

A chest X-ray revealed a para-cardiac speculated opacity. Viewing these features, a primitive neoplasm of the lung with metastasis to the vault was suggested.

The decision was to operate the patient to perform an excision of the cranial tumor. Perioperatively, the first step consisted of excision of the subcutaneous cystic tumor that was easily cleavable from the skin; the tumor was composed of very thin-walled chambers filled with yellowish, putrid, dense liquid. The second step consisted of the removal of the invaded cranial vault. An oval craniectomy from either side of the SSS was mandatory to obtain safe margins of 2 cm all around the tumor. The underlying dura mater was invaded by a friable reddish fleshy tumor. Durotomy was performed with flushing of the SSS; the upper wall was curetted and coagulated. Reconstruction consisted

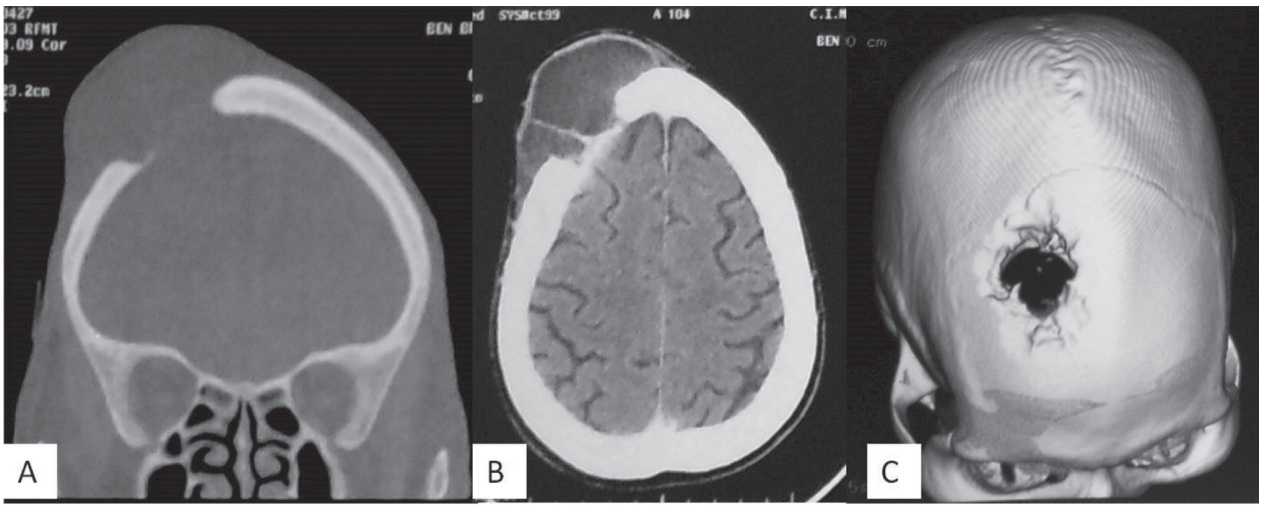

Figure 2. Preoperative cerebral CT scan: (A) bone (B) cerebral and (C) 3D reconstruction; a cystic multilobulated subcutaneous mass is present in the right frontal region with vault osteolysis.

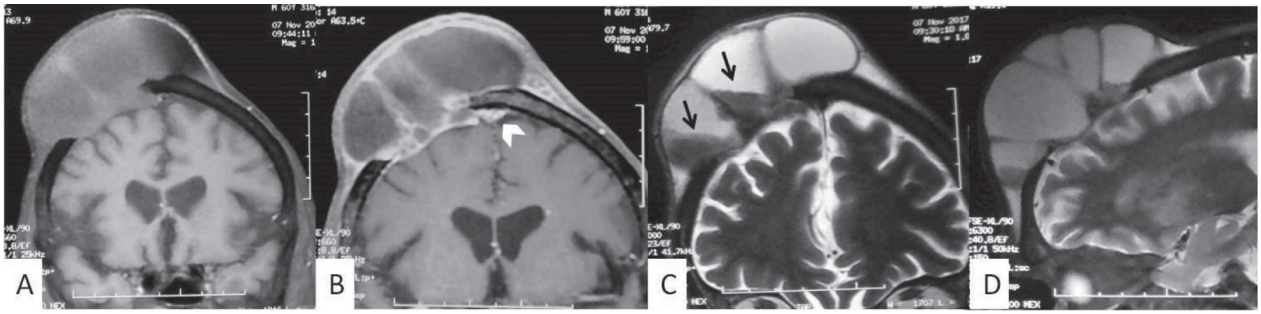

Figure 3. Magnetic resonance imaging: T1-weighted image (A), coronal T1-weighted image with gadolinium (B), coronal T2-weighted image (C), and sagittal T2-weighted image (D), showing a multilobulated cystic mass with liquid-liquid levels (arrow) responsible for bone lysis, invading the dura mater and coming into contact with the superior sagittal sinus, which remains permeable (arrowhead).

of a tight duraplasty using the epicranium, and a cranioplasty with acrylic cement. Excessive skin was removed for proper closure of the skin (Figure 4).

The postoperative course was uneventful. A control CT scan did not show any tumor residue or complication of the procedure. The histological examination confirmed the diagnosis of an MPTT (Figure 5). Afterward, the patient was referred to the Department of Oncology. A chest CT scan confirmed the presence of a lung tumor. A fibroscopic biopsy was attempted but without success. Otherwise, no other metastatic localizations were found. He underwent radiotherapy and chemotherapy (Cisplatin and 5-Fluorouracil). Six months later, no recurrence or evolution was noticed. The lung image remained stable.

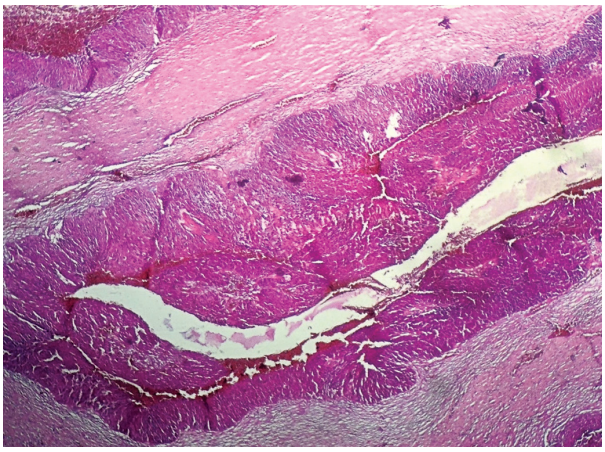

Figure 5. Pathological examination: HE × 20: Trichilemmal keratinization associated with an infiltrating carcinomatous squamous proliferation organized in lobules and clumps.

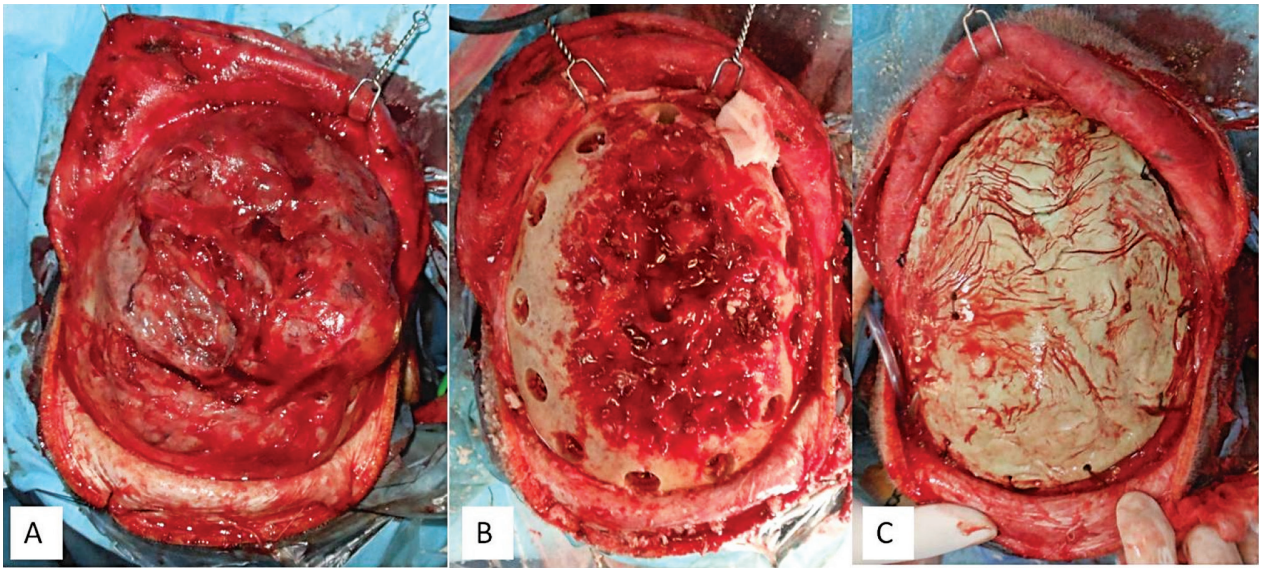

Figure 4. Photographs at different times of surgery: tumor resection (A), craniectomy of the invaded vault (B), repair time: cranioplasty (C).

DISCUSSION

MPTTs are extremely rare cutaneous tumors originating from the outer root sheath of hair follicles. Only about 50 cases have been reported in the 50 years since it was first described [5]. These tumors occur mainly in the scalp, eyelids, neck, and face of women over the age of 40 [6,7]. Macroscopically, they are exophytic, often ulcerated, polypoid, sometimes nodular, and dyskeratotic. Histologically, MPTTs show significant nuclear atypia, marked cellular pleomorphism with atypical mitoses, as well as dyskeratotic cells and stromal infiltration [4]. MPTTs can emerge de novo, but they most often occur within a pre-existing benign PTT, with the malignant transformation being in a stepwise manner: adenomatous, epitheliomatous, and then carcinomatous [6,8]. In fact, there is no distinctive immunohistochemical marker to detect this malignant transformation [6]. There is some evidence that this degenerescence maybe suggested by some features, including rapid growth, surface ulceration, local invasion, necrosis, multiple mitoses, and marked cytonuclear atypia, although the latter two are also present in benign PTTs [3,9,10]. Some authors retain infiltration of the stroma as an element of confirmation of the malignancy [11].

The actual incidence of MPTTs is not known because of their rarity and their misclassification as other malignant skin tumors. Differential diagnoses of MPTT are mainly squamous cell carcinoma, basal cell carcinoma, and pilomatrix carcinoma [2,12,13].

The principle for treatment of MPTT without metastasis is wide surgical excision with a margin of 1 cm in normal tissue [2,12]. To our knowledge, there is only one other case of MPTT reported in the literature with endocranial extension and lung metastasis at the time of diagnosis [4]. Very few other cases presented endocranial extension and lung metastases after primary surgery for non-invasive MPTT, demonstrating the relapsing and aggressive nature of these lesions [3,14]. The metastatic rate of MPTTs is estimated to be 25% [5]. Endocranial extension and distant metastasis have been a hindrance to the surgical decision in all of these cases. Treatment in these cases was palliative, associating chemotherapy (six cycles of cisplatin and 5-fluorouracil) and radiotherapy [3-5,10,14]. The patients died two months after the end of palliative treatment [4]. For the others, there was no sufficient hindsight. There is indeed no consensus, and the management of MPTT remains empirical [3]. Following the recommendations of El Benaye *et al.* [3], we attempted a radical excision with a wide margin of safety (2 cm) given the large size of the lesion and the high presumption of aggressiveness. Adjuvant treatment with radiotherapy and chemotherapy was administered with a good outcome at six months. Long-term follow-up of this case and other similar cases would allow comparison of patient survival and would possibly put surgery back in the therapeutic arsenal for invasive and metastatic MPTT.

CONCLUSIONS

MPTT associated with intracranial extension and lung metastasis is an extremely rare pathological condition. At this stage of the disease, palliative treatment is generally recommended. We propose radical surgery associated with adjuvant treatment. Long-term follow-up is necessary to judge the contribution of this intervention to prognosis and survival.

Conflict of interest: None declared.

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