SAS Journal of Surgery
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

Surgery

A Rare Tumor of Scalp - Giant Multiple Cellular Histiocytofibroma

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DOI: 10.36347/sasjs.2024.v10i05.004 | **Received:** 06.08.2023 | **Accepted:** 14.09.2023 | **Published:** 09.05.2024

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Abstract **Case Report**

Histiocytofibroma or dermatofibroma is a rare benign cutaneous tumor present preferentially in the dermis. Histiocytofibroma is most frequent benign tumor, but some specific morphological forms may present local recurrences and can cause lymph node and visceral metastases. The trunk and limbs represent the most classically described topography, other locations are known like face and scalp. Histiocytofibroma is most often present as a unique lesion: papule or nodule, giant form is not common and it is characterized by a large tumor volume (5 cm or more). The cellular histological variant is a rare subtype. We report a case of a 56-year-old male patient with no particular past medical history, who presented with a fronto-parietal cutaneous large tumor evolved for 2 years, gradually increasing in volume, measuring 7 cm in diameter, with regular contours, flesh colored, and peripheral vascularization on the surface of the lesion, it was firm and adherent to bone plane at palpation, along with seven other small lesions, the largest of which was 2 cm. The CT scan did not show any bone invasion. Surgical management consisted of large excision with 1 cm free margins. histological and immunohistochemical findings were in favor of cellular histiocytofibroma. The substance loss was covered by skin graft after complete budding. A 26 months follow up results were good without recurrence.

Keywords: Histiocytofibroma, Rare, Excision.

INTRODUCTION

Histiocytofibroma (HCF) is a common cutaneous tumor present preferentially in the dermis. This lesion also known by many other names including the most common dermatofibroma. The trunk and limbs represent the topography that is most classically described, other locations such as the face and scalp are known. It is most often presented as a papule, nodule or plaque, but giant forms too have been described which is most often unique, and the multiple forms are rare. The histological cellular variant is associated with the risk of local recurrence and most often this type is large. We report a rare case of a 56-year-old male patient with a rare form of giant, multiple and histological cellular fronto-parietal HCF.

A large surgical resection has been performed. Follow up was good without sign of recurrence.

CASE REPORT

This is a case of a 56-year-old patient, operated twice for a scalp tumor (documents not available), without other pathological history, which presented a mass of the left fronto-parietal region evolving for 2 years, gradually growing in size, associated with multiple smaller lesions. The clinical examination found a stable patient with a left frontal-parietal large tumor measuring 7x5x4cm, with regular contours, flesh colored, and peripheral vascularization on the surface of the lesion, at palpation, the lesion was painless, firm in consistency and adherent to the bone plane, along with seven other small lesions, the largest of which was 2 cm, (Figure 1). No cervical lymphadenopathies were clinically palpated.

The CT scan revealed the presence multiple spontaneously hypodense formations of variable sizes at the level of the frontal scalp, the largest of which measuring 68x47mm without any bone invasion. An ultrasound of cervical lymph nodes was performed,

returned without abnormalities, a cut biopsy came back in favor of Darier–Ferrand dermatofibrosarcoma. A surgical excision was performed including the seven small lesions with wide margins (Figures 2 and 3). The pathological and immunohistochemical study returned in favor of cellular HCF. The tumor cells were found to contain CD34 and AML to some degree. However, no S100 or CD68 or desmine were found. The loss of substance was covered after three weeks by a split thickness skin graft after budding (Figures 4 and 5). A 26 months follow up results were good without recurrence.

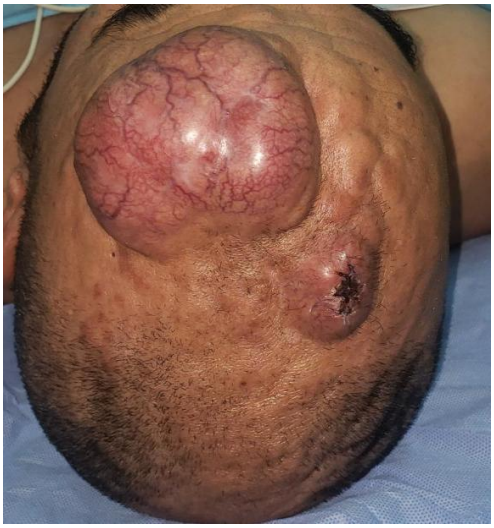

Figure 1: Clinical image of the tumor

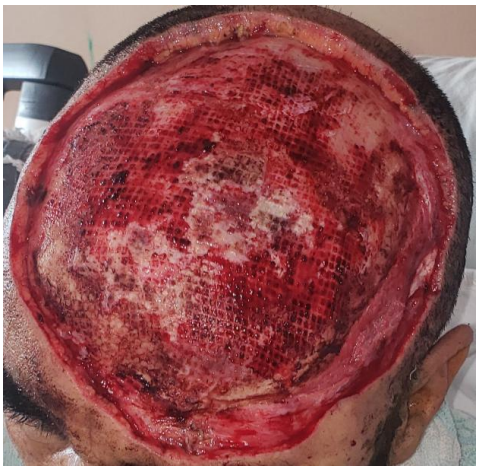

Figure 2: Scalp appearance after surgical removal

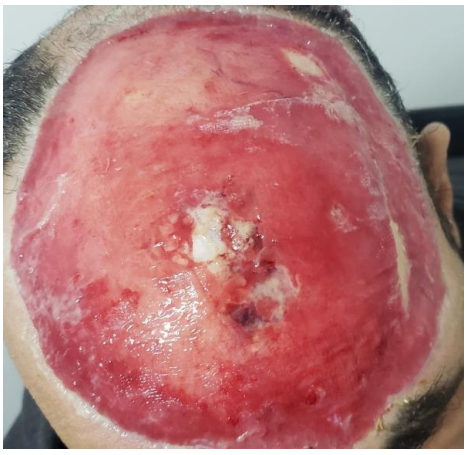

Figure 3: Clinical appearance after budding

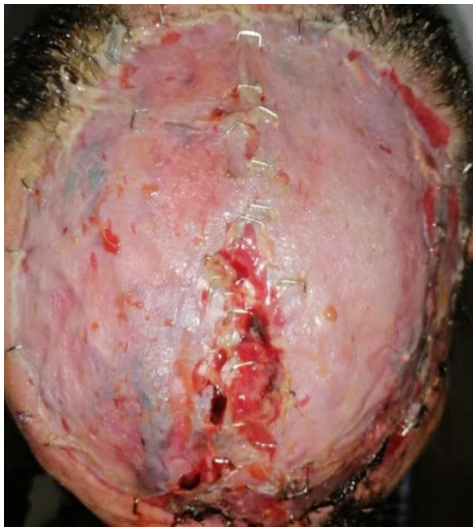

Figure 4: Clinical appearance after skin graft 3 weeks later

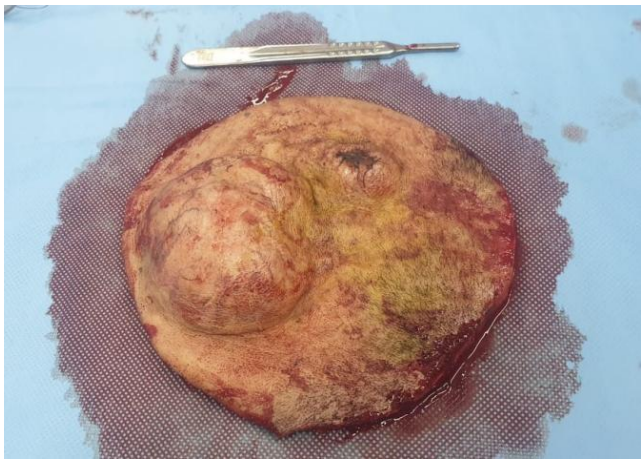

Figure 5: Surgical specimen after resection

DISCUSSION

HCF or dermatofibroma is a cutaneous tumor present preferentially in the dermis. HCF is most frequent benign tumor, but some specific morphological forms may present local recurrences and can cause lymph node and visceral metastases [1]. HCF can occur at any age but most commonly affects the young age and middle age adults (20–40 years) with female predominance [2]. The trunk and limbs represent the most classically described topography. The other known locations are the face and scalp [3]. The lesion grows fast in the early phase, but its size stabilizes over time and is often sub-centimetric (91.8%). HCF is often asymptomatic, however can be sensitive (20.5%), painful (11.5%) or itchy (58.2%), leading patients to consult the dermatologist [4, 5]. HCF is most often seen as a papule, nodule or plaque, but giant forms have also been described. The giant form is very rare and is characterized by a large size (5 cm), which may cause suspicion of malignancy [6, 7]. HCF is most often unique, multiple forms have been described, but in particular contexts, such as immunosuppression, autoimmune diseases, blood diseases, metabolic diseases and pregnancy [8]. The classic form of HCF (80% of cases) is defined in the Classification of WHO Skin Classification of Skin Tumors (2018) as a papule or a nodule composed to varying degrees of fibrohistiocytic cells, of macrophages and coarse collagen [9]. The cellular histological variant is a rare subtype, less than 5%, this variant is more common in humans. The cellular form (2.1% of HCF) affects both men and women with a median age of 32 years. The topography of this variant is identical to the conventional HCF. This type of HCF is often larger and has cellular density with a proliferation in bundles with fusiform cells, “plump” only in the classical form, but without atypia [10]. This form is associated with a higher risk of local recurrence and is most often large in size. Cellular HCFs recur locally in 10% of cases. The predictive factors of local recurrence are size greater than 1 cm and positivity of margins during the initial excision [6]. The most serious significant differential diagnosis is Darier–Ferrand dermatofibrosarcoma, immunohistochemistry can redirect the diagnosis [11]. Treatment is usually surgical removal and monitoring for high-risk histological variants [12] as in the case of our patient.

CONCLUSION

Histiocytofibroma also known as dermatofibroma is a common cutaneous tumor that evolves preferentially on trunk and limbs, other locations such as the face and scalp are known. Histiocytofibroma is often asymptomatic and it is most often seen as a papule, nodule or plaque, but giant forms too, have been described. Histologically, the cellular variant is a rare subtype.

The treatment is based on surgery followed by a regular follow-up especially in high risk variants.

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