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### RESEARCH ARTICLE

#### CUTANEOUS EPITHELOID HEMANGIOENDOTHELIOMA: A RARE TUMOR

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#### Abstract

Epithelioid hemangioendothelioma is a rare, slowly unfavorable vascular tumor. It is considered to be a malignant tumor. Localization in the skin is very rare and often part of a multi-systemic disorder. It has the potential for local recurrence, lymph node metastasis and at a distance and it's not frequently accompanied by death. We report the observation of a 57-years-old patient, who is presented with an epithelioid hemangioendothelioma of the left buttock with a 1-year follow-up without recurrence.

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#### Introduction:-

Epithelioid hemangioendothelioma (HHE) is a rare mesenchymal tumor of vascular differentiation, which develops, like angiosarcoma, to caricature endothelial cells. However, its histological aspect is particular because tumor cells adopt an epithelial cell morphology, hence the term "epithelioid" (Weiss and Enzinger, 1982). Currently, it is considered a malignant vascular tumor. HEE mainly occurs in the bone, lung, and liver. Skin localization is rare. We report the observation of a 57-years-old patient, who is presented with a cutaneous epithelioid hemangioendothelioma of the left buttock with a 1-year follow-up without recurrence.

#### Case report:-

Mrs. H.B 57-years-old, with no notable pathological history, is presented for a nodular lesion in the left buttock evolving for 3 years, gradually increasing in size and evolving in a context of conservation of the general state. The clinical examination objectified a bulky mass of 10 cm of the long axis, ulcero-necrotic, with serum outcome, slightly painful on palpation, fixed to the deep plane (Fig 1). The rest of the clinical examination was without particularities: no other associated lesions or palpable lymphadenopathies.

A biopsy was done revealing an aspect of malignant neoplastic process evoking an epithelioid hemangioendothelioma. A large excision was performed (Fig 2) associated with an immediate direct closure (Fig 3). An anatomopathological study of the excision piece showed a hyperplastic epidermis. The reticular dermis had a fibrous appearance and was the site of a cell proliferation made of cells with abundant cytoplasm, epithelioid appearance and regular nucleus with an intracytoplasmic vacuole. The suspected vascular nature was confirmed by an immunohistochemical study. These cells had an antigenic profile of endothelial cells: CD31 +, CD34 +. In conclusion, it was an epithelioid endothelial cell proliferation realizing the appearance of an epithelioid hemangioendothelioma. The extension assessment including a thoraco-abdomino-pelvic scanner did not show any secondary hepatic or pulmonary localizations. The postoperative follow-ups were simple. With a one-year follow-up, we didn't note any locoregional recurrence or distant metastasis.

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**Discussion:-**

Epitheloid hemangioendothelioma (HEE) is a rare tumor of vascular origin. The term HEE was originally proposed by Weiss and Enzinger in 1982 (Weiss and Enzinger, 1982) to refer to a heterogeneous group of vascular tumors with clinical, histological and progressive features that are intermediate between hemangiomas and angiosarcomas. Its etiology is to date unknown. According to the latest classification from the International Society for the Study of Vascular Anomalies (ISSVA) in 2018, HEE is classified as one of the malignant tumors. It represents 1% of vascular tumors. In more than 60% of cases, it initially affects only one organ, mainly the liver (34%), the bones (21%), or the lungs (19%) (Lau and al, 2011). The localization in soft tissue is rare. Our observation is particular by its cutaneous localization. HEE has no predilection for the skin, 23 cases of HEE localized at this level have been reported (Fenniche and al, 2004). It's observed in middle-aged adults, rarely in childhood and both in men and women (Bardouni and al, 2014). HEE is characterized by clinical latency with a consultation time of up to 10 years or more in one third of patients (Enzinger 2001, Fletcher 2002). Typically, patients with HEE present infiltrating masses, uninodular or multinodular, of dermal or subcutaneous site, the growth of which takes place very slowly.

These masses sit in most cases at the distal ends. The head, neck, thorax and abdomen have also been reported as sites of origin of the tumor (Bardouni and al, 2014). In our case, it was a localization in the buttock hence the originality of our observation. HEE is defined histologically as an angiocentric vascular tumor. The tumor cells are epithelioids, more rarely spindle shaped, with an atypical nucleus, often vesicular, with a small nucleolus, rarely in mitosis and the cytoplasm sometimes contains a vacuole containing hematoma (vascular differentiation at the cellular level). These cells are arranged in spans, cords or islets, in a myxoid or hyaline background. The intracytoplasmic vacuole can be large and compress the nucleus, creating cell appearances in a kitten ring. Territories of necrosis, calcifications/ossifications, giant multinucleated cells, hemosiderin deposits and an inflammatory infiltrate can be seen (Chenard and al, 2016). The immunohistochemical study is a great help to confirm the endothelial nature of HEE. Indeed, the positivity of the labeling of tumor cells with anti-CD34, anti-factor VIII and anti-CD31 (more specific but more discreet) antibodies enables the diagnosis to be made (Hajji and al, 2002). In a third of cases, one can observe a positivity for cytokeratins. A cytogenetic study of 2 cases of HEE, localized in the liver and soft tissues, made it possible to highlight the presence of a translocation between chromosomes 1 and 3 (t (1; 3) (p36.3; q25) which seems to be potentially characteristic of HEE (Hannachi 2005, Mendlick and al 2001) Unfortunately, we were unable to carry out the cytogenetic study in our context. Multifocal localizations of HEE are rare and have a poor prognosis. The extension must be carried out in all cases. In our case, no extra-cutaneous localization was found.

Although most of these tumors behave indolently, 20-30% of cases have a metastatic course and 15% of patients die from this condition. Cases of partial spontaneous regression are described (Sardaro and al, 2014). Overall survival is 73% at 5 years (Antonescu, 2014). The prognosis is worse in the case of multi-organ localization. Morphological criteria were proposed by Deyrup and al (Deyrup and al, 2008) to separate HEE into two groups, low and high risk (criteria not valid for multifocal and visceral presentations): tumor size > 3 cm and > 3 mitoses / 50 HPF. Patients whose tumor has these characteristics have a 5-years survival rate of 59%, compared to 100% survival for patients whose tumor doesn't have these characteristics.

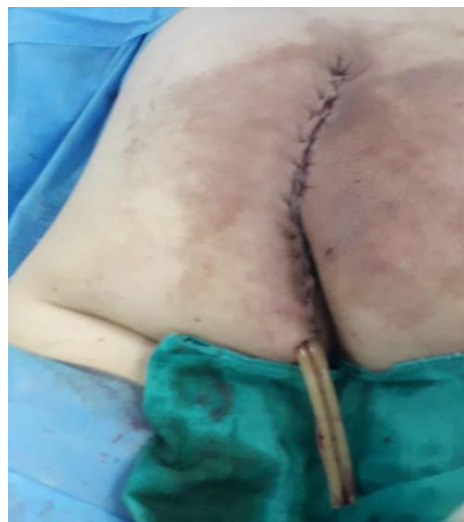
The treatment of HEE isn't well codified. Surgery is particularly difficult because of the poor peripheral delimitation of the lesion and the diffuse infiltration of the muscles, tendons and underlying vasculo-nerve structures. Thus, the resection must be as wide as possible (Dhawan and al, 2007). This is what has been achieved in our patient. Recombinant interleukin 2, a derivative of T-cell lymphokines, has been successfully tested (Dhawan and al, 2007). Adjuvant radiotherapy is especially indicated in multicentric forms and seems to be effective. Multi-drug therapy used in aggressive multifocal forms has no clearly demonstrated efficacy (Hajji and al, 2002).



**Figure 1 :** ulcero-necrotic mass in the left buttock.



**Figure 2 :** 2cm margins of excision



**Figure 3 :** Excision of the tumor and immediate direct closure.

**Conclusion:-**

HEE in soft tissue is a rare malignant tumor of vascular origin. It thus involves a risk of local recurrence, lymph node invasion and metastasis from a distance. Its clinical expression is generally rough. The diagnosis is based on the histological and immunohistochemical study. Therapeutic management remains poorly codified. However, the treatment is essentially surgical and must be as conservative as possible.

**References:-**

1. Ahmed Bardouni et Al, Hémangioendothéliome épithélioïde de la cuisse: à propos d'un cas, The Pan African Medical Journal - ISSN 1937-8688, 2014
2. Antonescu C. Malignant vascular tumors - an update. Mod Pathol 2014; 27:S30-S8
3. Deyrup AT, Tighiouart M, Montag AG, Weiss SW. Epithelioid hemangioendothelioma of soft tissue: a proposal for risk stratification based on 49 cases. Am J Surg Pathol 2008;32:924-7
4. Dhawan SS, Raza M. Spindle cell hemangioendothelioma. Cutis. 2007 Feb;79(2):125-8
5. Enzinger FM. Soft Tissue Tumors, fourth edition. 2001. Mosby. St. Louis.
6. Fenniche S, Benmously R, Marrak H, Khayat O, Ben Ammar F, Debbiche A, et al. Hémangioendothéliome épithélioïde de cutane'. Ann Dermatol Venerol 2004;131:818-21
7. Fletcher CDM. Tumors of soft tissue and bone Pathology and Genetics. 2002. Lyon. IARC Press
8. Hajji N, Kallel MH, Sellami M, Ayadi K et al. Hémangioendothéliome osseux multicentrique. A propos d'un cas. Rev Chir Orthop. 2002; 88(1): 78-81
9. Hannachi Sassi S. Soft tissue epithelioid hemangioendothelioma: a case report. Rev Chir Orthop Reparatrice Appar Mot. 2005 Nov;91(7):671-5
10. Lau K, Massad M, Pollak C, Rubin C, Yeh J, Wanq J, et al. Clinical patterns and outcome in epithelioid hemangioendothelioma with or without pulmonary involvement: insights from an internet registry in the study of rare cancer. Chest 2011;140:1312-8.
11. Marie-Pierre CHENARD et al, DIFFICULTES DIAGNOSTIQUES SUR BIOPSIES MAMMAIRES, HISTOSEMINAIRE CARREFOUR PATHOLOGIE 2016
12. Mendlick MR, Nelson M, Pickering D, Johansson SL et al. Translocation t(1;3)(p36.3;q25) is a nonrandom aberration in epithelioid hemangioendothelioma. Am J Surg Pathol. 2001 May;25(5):684-7
13. Palsson B. Epithelioid hemangioendothelioma. Acta Oncol. 1999;38(5):659-61
14. Piard F. Hémangioendothéliome épithélioïde. Bulletin de Division Française de l'AIP no 41 ; 2005.
15. Sardaro A, Bardoscia L, Petruzelli MF, Portaluri M. Epithelioid hemangioendothelioma : an overview and update on a rare vascular tumor. Oncol Rev 2014;8:259.
16. Weiss SW, Enzinger FM. Epithelioid hemangioendothelioma a vascular tumor often mistaken for a carcinoma. Cancer. 1982 Sep 1;50(5):970-81.