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Case Report

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PSEUDOMYOGENIC HEMANGIOENDOTHELIOMA OF THE KNEE- A CASE REPORT

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ABSTRACT

Pseudomyogenic hemangioendothelioma also known as PMHE are a rare vascular tumor discovered recently which affects young subjects and particulary their limb. The clinical presentation in early stages is non specific with the possible appearance of dermal or subdermal nodules. MRI is the gold standard exam that helps guide the diagnosis made by the biopsy. The treatement of PHME is wide surgical resection with amputation at the last resort.

INTRODUCTION

Pseudomyogenic hemangioendothelioma (pmhe) is a rare and unusual tumor of loud tissue, first described in 1992, and classified in 2003 as a vascular tumor by WHO. It particularly affects young subjects with predilection on the lower limb. The diagnosis is histhopathological and treatment is surgical. We report the case of a patient being treated in our training.

CASE REPORT

A 63 year old male patient, originally from and living in rabat the capital of morocco, with a history of chronic smoking at the rate of 40 pack-years. The history of the disease begins in 2016 with the appearance of a non-itchy erythematous plaque internal to the right knee, that became a few weeks later hard and containing a small polypoid formation (figure 1).

The patient then received a standard x-ray of the knee not showing any bone damage. An MRI was requested which revealed a mass of the soft parts of the internal compartment of the knee, hyposignal in T1 and hypersignal in T2 measuring 82x42 mm in diameter (figure 2).

The patient first underwent a percutaneous biopsy which returned negative. The patient was then taken to the operating room where he underwent a wide surgical resection. The remaining anatomical piece was sent to the pathology laboratory where the PMHE diagnosis was made. The evolution of the desease was marked by tumor recurrence on several occasions and the performance of multiple surgical resections, that caused the loss of skin substance.

Faced with a recurrence in 2020, a thigh amputation was performed. One year after the amputation, the patient's condition has progressed well without any recurrence.

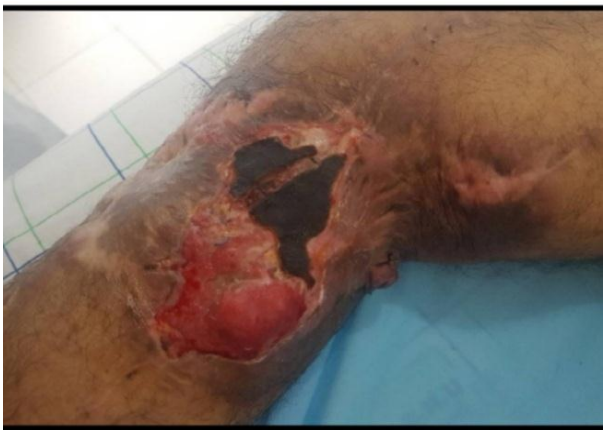

Figure 1: Clinical aspect of the knee.

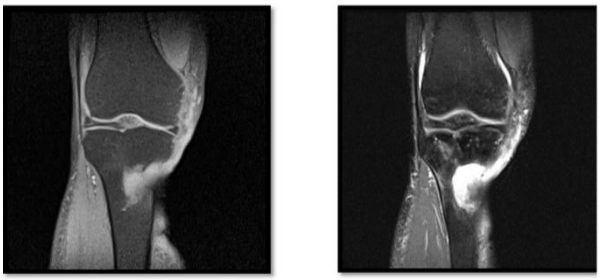

Figure 2: MRI of the knee with hyposignal and hypersignal images in T1 and T2 respectively.

DISCUSSION

PMHE are rare and unusual soft tissue tumors described in 1992 by Mirra et al.[1] In 2003, Bellinger and Andall

reported 7 similar cases and referred to these tumors as sarcoma epithelial hemangiothelioma. Hornick and Fletcher published a study confirming the vascular origin of these tumors.[2] They affect men more than women with an average age of 30 years and with a predilection for the lower extremity 59 %, upper extremity 18%.

The clinical presentation is nonspecific. Pain is the most frequent symptom, inflammatory in nature, localized, increasing in intensity with the progression of the disease, with possible appearance of dermal or subdermal nodules sometimes having a multifocal character implicating more than one tissue plane: superficial soft tissue, deep soft tissue, muscle tissue, bone tissue.[3]

MRI is the exam of choice for exploring soft tissue pathologies. These lesions appear as a T1 hypointense and T2 hypersignal image.[4] It allows a better evaluation of the tumor lesion and guides the diagnosis.

The latter is confirmed by biopsy. It has been mentioned by several studies that percutaneous biopsies do not lead to a definite diagnosis of this pathology and therefore resort to surgical biopsy being mandatory in order to obtain a diagnosis.[5]

The treatment is not well established by the literature. However, the most important element to beat this desease is conservative care. In rare cases, especially aggressive PMHE, chemotherapy and radiotherapy have been indicated and have given relevant results. Wide excision is the surgical standard; the tumor is removed with a healthy tissue margin of 1 to 2cm over its entire surface.[6] The spontaneous progression of this pathology is generally towards an increase in the size of the lesion with the risk of compression of the surrounding structures. The main prognostic factors for predicting the recurrence of this pathology are age, gender, size of lesions.[7-8]

CONCLUSION

Pseudomyogenic hemangioendothelioma is a relatively rare vascular tumor described for the first time in the early 1990. The diagnosis is difficult and relies on performing multiple biopsies. The treatment is mainly surgical and the prognosis is good according to the literature.

Conflicts of interest

The authors declare no conflict of interest.

Authors' contributions

All authors contributed to the care of the patients and the writing of the manuscript. All have read and approved the final version of the manuscript.

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