

## Reactive angioendotheliomatosis: A study of one case and review of literature

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### Summary

Reactive angioendotheliomatosis is a rare lesion that usually occurs in patients with systemic diseases which have renal failure and are treated by haemodialysis. It is a benign pathology with an anatomopathological diagnosis and little codified treatment. We report here the case of a 34-year-old patient with right hand, left hand and right foot localization and without any systemic disease.

**Keywords:** Angioendotheliomatosis; papule; granulomatous.

### Resume

L'angioendothéliomatose réactionnelle est une lésion rare qui apparaît généralement chez les patients atteints de maladies systémiques qui ont une insuffisance rénale et sont traités par hémodialyse. Il s'agit d'une pathologie bénigne dont le diagnostic est anatomopathologique et le traitement peu codifié. Nous rapportons ici le cas d'un patient de 34 ans à localisation main droite, main gauche et pied droit et en dehors de toute affection systémique.

### Introduction

Reactive angioendotheliomatosis is a rare lesion [1-3]. It usually occurs in patients with systemic diseases who have renal failure and are treated with haemodialysis [4-6]. We report here an isolated granular reactive angioendotheliomatosis of the extremities of the limbs in the absence of systemic disease.

### Observation

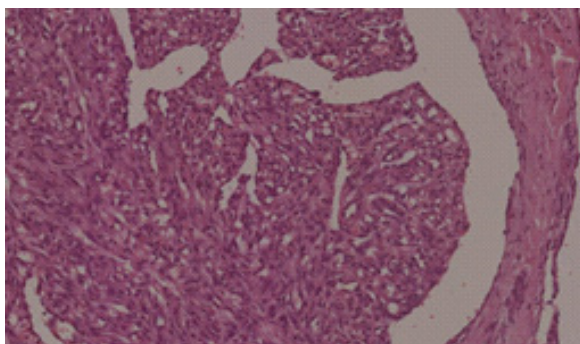
This is a 34-year-old patient with no previous pathological history who was seen in consultation for painless granular swellings on both hands and the right foot, that had been developed for 2 years. The clinical examination revealed: a good general condition and good hemodynamic state. Locally, he had multiple papules on both hands and the right foot. The

lesion was of variable diameter, rosaceous in colour, painless and of different ages (**Figure 1**). On examination, the diagnosis of Kaposi's sarcoma was suggested. A biopsy was performed. Anatomopathological examination of two fragments revealed a benign vascular accral proliferation of nodular configuration involving both the superficial and deep dermis arranged in intravascularly growing glomeruloid nodules without atypia compatible with reactive angioendotheliomatosis (**Figure 2 and 3**). No management was conducted in the patient as the patient was lost to follow-up.

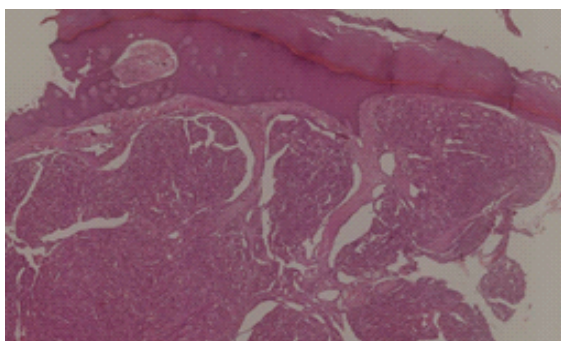


**Figure 1:** Lesion seen in the hand.

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**Figure 2:** Anatomopathology picture.



**Figure 3:** Anatomopathology picture.

## Discussion

Angioendotheliomatosis is a rare, benign condition with polymorphic clinical manifestations ranging from macules to plaques or nodules that may be purpuric, ulcerated or scleroderma-like [1-7]. It is often associated with systemic diseases [8]. Some cases have been described in patients with cryoproteinaemia [7]. One hypothesis to explain the association with these diseases is that they are all characterised by high oxidative stress, which is also increased by haemodialysis and facilitates vascular [7]. Furthermore, in these diseases, hypoxia and skin reperfusion processes can occur which locally increase the concentration of oxygen free radicals, which are known to be a potent angiogenic factor [1, 2]. It is characterised by a specific histological pattern of proliferating dermal capillaries, the lumens of which are obliterated by proliferating endothelial cells [9-11]. Two anatomopathological types are frequently found. These are intravascular reactive angioendotheliomatosis and diffuse reactive angioendotheliomatosis [2]. The second form, which is the one described in our clinical case, may be accompanied by cavernous dilatations, intravascular thrombi and, in some cases, epithelioid endothelium. Endothelial atypia is an unusual phenomenon although it is found in some patients. This atypia was not found in our case [2].

The differential diagnosis is most often made with kaposi's sarcoma [6, 9]. It is a benign pathology [8, 11]. The diagnosis is anatomopathological [7]. The treatment is not well codified. It is based on the management of risk factors for atheroma and on clean surgery. The efficacy of systematic corticosteroid therapy, effective anticoagulant and revascularisation have been reported [12].

## Conclusion

Angioendotheliomatosis is a rare and little-known condition. The diagnosis can be made by pathological examination alone.

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