CASE REPORT

Aneurysmal dermatofibroma mimicking both clinical and dermoscopic malignant melanoma and Kaposi’s sarcoma

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Abstract

Aneurysmal dermatofibroma (AD) or aneurysmal fibrous histiocytoma (AFH) is a relatively rare form of histiocytoma representing less than 2% of total cases. It shares many clinical and dermoscopic similarities with skin tumors, especially malignant melanoma and Kaposi’s sarcoma, which may make differentiation problematic. We report the case of a 53-year-old man, who presents with a black nodular tumor with increased consistency, edges infiltrated from the surface to depth, spontaneous and sensitive to touch that shows rapid growth in the last three months. Dermo-scopically, the central region consists of intricate areas colored in red, violet, blue-white, and black. On the periphery stand two rings, centripetally white and peripherally pigmented, with an abundance of polymorphic capillaries. To clarify the diagnosis, the lesion was widely excised and histopathological examination was performed, which revealed immunophenotypical tumor cells negative for HMB-45 and S100 and numerous CD68 macrophages between tumor cells. This aneurysmal fibrous histiocytoma shows both clinical and dermoscopically discrete differential criteria, which are not specific and that make it difficult to distinguish from malignant melanoma and Kaposi’s sarcoma, and required performing histopathology and immunohistochemistry.

Keywords: aneurysmal dermatofibroma, aneurysmal fibrous histiocytoma, malignant melanoma, Kaposi’s sarcoma, dermoscopy.

Introduction

Dermatofibroma or fibrous histiocytoma (FH) is the most common fibrous tumor found in the dermis. In common forms, it is easily diagnosed and differentiated clinically and dermoscopically from other skin tumors because of dermoscopy criteria defined by Zaballos et al. [1]. The presence of a pigment network and central white patch allows the experienced examiner to diagnose with certainty, and a conservative therapeutic approach to be set [2]. In practice, there are often situations in which clinical and dermoscopic specific features present in conjunction with the absence of historical data and evolution prevents follow-up. In these situations, the correct diagnosis requires microscopic examination and immunohistochemistry [3]. Aneurysmal fibrous histiocytoma (AFH) represents less than 2% of all FH [4]. AFH was first described by Santa Cruz and Kyriakos, in 1981, as a variety of cutaneous fibrous histiocytoma [5]. Unlike the classic version of FH, the aneurysmal variant may be more elevated, with a color ranging from blue to black and a softer consistency, cystic [5, 6], growing faster than the classical FH and may be accompanied by pain. These particular features distinguish it from the classical form but draw it closer to other diagnoses including evolving tumor formation and a more severe prognosis, as malignant melanoma (MM) [7, 8] and Kaposi’s sarcoma (KS) [9, 10].

The clinical picture of AFH lends itself to confusion in current practice. Different therapeutic attitudes are required in this case of benign tumor as opposed to MM and KS. In these cases, additional methods of investigation are mandatory before management decisions are made. Some authors have tried to define a specific distinguishing dermoscopic appearance [11].

Patient, Methods and Results

We report a case of a 53-year-old male from an urban area, presenting with a lesion on the posterior thorax in the scapular region. The 12 mm diameter lesion had a nodular formation with a black surface covered with fine scales, high consistency, infiltrated edges from surface to depth with a red/purple glow, spontaneous appearance and sensitive to touch (Figure 1a). The patient reports the lesion onset a few years previously, with three months of rapid growth. With size, increasing the lesion became sensitive to touch. Patient reported no previous trauma to the area involved, but the area is exposed to chronic irritation caused by clothing and wearing a shoulder bag. Dimple sign was absent. General clinical examination did not reveal the presence of regional adenopathy. Laboratory tests reveal no significant changes. Dermo-scopically (Figure 1b), we observed in the centre of the lesion an unstructured aspect looking like a constellation, consisting of red-purple and white-blue areas. This area, along with the black areas on the periphery, resembled the blue-white veil characteristic of malignant melanoma. Two rings surround the central region, a white interior ring reminiscent of the central area of fibrous histiocytomas, and a peripheral light-brown pigmented circle, specific also to FH. Throughout the nodular lesion, we observed
a scaly surface. Corresponding to the infiltrated area surrounding the lesion, we noticed numerous polymorphic capillaries.

Figure 1 – (a) Clinical aspect; (b) Dermoscopy aspect.

Malignant melanoma could not be excluded, so wide excision of the lesion with a malignant melanoma specific safety zone of 2 cm was performed.

On microscopy was observed an intact keratinized stratified squamous epithelium, showing a poorly defined nodular tumor proliferation in the middle and deep dermis. The papillary dermis was unaffected. Flattening rete ridges and a thinning of spinosum layer was noticed in the superficial epithelium. At the edge of the skin flap, acanthosis and basal layer hyperpigmentation were observed. In the centre of the lesion, there were numerous large vascular spaces, forming irregular cleft-like spaces without endothelium. Some vascular spaces were filled with erythrocytes; others had disrupted walls with interstitial hemorrhage. Around the periphery of the lesion, there were extensive deposits of dark-brown pigment, both extracellular and intracellular. At the periphery of the lesion, a classic tumor pattern was observed: solid, with proliferation of fibroblasts and histiocytes, in a rich connective vascular stroma, without mitoses. The hypodermis was tumor-free and the perilesional skin appendages were microscopically normal.

Special stainings and immunohistochemistry

Perls’ Prussian blue staining confirmed the presence of extracellular and intracellular hemosiderin deposits.

Immunohistochemically, numerous CD68 macrophages between tumor cells were observed. The tumor cells were negative for HMB-45 and S100. Vascular endothelium presented positively for CD31 and CD34.

Pathological diagnosis was aneurysmal fibrous histiocytoma (AFH), excised in safety surgical margins.

The clinical and dermatoscopic exams performed regularly over the past 36 months revealed no local recurrence or metastasis.

Figure 2 – (a) Microscopic aspect, Hematoxylin–Eosin staining, ×20; (b) Hemosiderin deposits, Perls’ Prussian blue staining, ×20; (c) CD68+ macrophage, CD68 immunostaining, ×100; (d) CD34+ vascular endothelium, CD34 immunostaining, ×40.

Discussion

Particularities of the case which further complicated diagnosis are the presence of infiltrated edges from surface to depth and intense black color, the presence of which is theoretically possible in FH, but is still rare in practice. The red-purple perilesional inflammatory halo
was also abnormal. The dimple sign or Fitzpatrick’s sign [12] is not present in our patient, probably due to perilesional inflammation. Zaballos et al. define a characteristic pattern of FH: peripheral pigmented network and a central white patch [13], corresponding with classic FH. In hemosiderotic fibrous histiocytoma and in AFH, the same authors describe the presence of a central homogeneous area toned red or blue, delicate white network pigmentosa and peripheral dotted, linear, comma-shaped and dilated vessel appearance, as well as isolated irregular vessels and a scaly surface [14]. We have highlighted in the central area an unstructured constellation-like aspect consisting of purplish-red and white-blue areas reminiscent of the malignant melanoma white-blue veil.

AFH is considered a particular form of histiocytoma defined by large, blood-filled spaces without endothelial lining. Hemosiderotic dermatofibroma is a variant composed of numerous small vessels, extravasated erythrocytes, and intra/extracellular hemosiderin deposits. Some authors consider aneurysmal histiocytoma as an advanced stage of hemosiderotic dermatofibroma, differentiated only by histopathology [13]. This particular structure is described by dermoscopy. The inner white ring is reminiscent of a central white spot, a characteristic of common dermatofibroma. The pigmented network at the periphery correlated with the long history of the lesion, much accentuated in the last three months since the injury became sensitive to the touch, allows us to postulate that the hemosiderotic dermatofibroma and aneurysmal dermatofibroma could be complications of a common dermatofibroma, probably induced by chronic local irritation caused by clothing and the chronic friction caused by the shoulder bag.

AFH clinical images sometimes resemble those of KS nodular form defined by red-brown dermo/hypodermic nodules, which can become painful in evolution. They can also appear similar to nodular MM, with a round and intensely pigmented smooth surface and a pigmented halo. Dermatoscopic examination of KS as described by Hu et al. highlights the presence of bluish-reddish coloration in 84% of cases, and multi-coloration in 36% [15]. This multicolored rainbow appearance does not differ greatly from the constellation aspect of our case. Another similarity is given by the presence of a scaly surface. Unlike KS, in AFH there is a peripheral double ring, white inside and pigmented outside, with polymorphic capillaries.

Relatively discrete aspects that distinguish AFH from KS and some MM, often do not allow certain diagnostic differentiation of the three conditions, and even for experienced dermatologists can be dermoscopic traps [13]. The implications associated with the development and more severe prognosis of MM and KS require excision of the lesion within safety margins, followed by histopathological examination and immunohistochemistry [16].

Histopathologically, although fibrous histiocytoma is the most common fibrous tumor in dermis, it remains a current topic because of the many clinical and microscopic forms. The identification of particular variants still represents a particular interest in daily practice, as mistakes in this regard can be life threatening. The most important differentiated diagnosis is with a melanocytic lesion: malignant melanoma or vascular lesion: Kaposi’s sarcoma. In both cases, the treatment of the patient and evolution of the lesion are significantly different from dermatofibroma.

Special stainings and immunohistochemistry can resolve this dilemma and produce the correct diagnosis. In aneurysmal dermatofibroma, hemosiderin deposits and many hemosiderophages are observed. Tumor cells types include fibroblasts and histiocytes, and vascular endothelial cells from the small vessels are positive for CD31 and CD34. The tumor cells are negative for HMB-45 and S100. Surface epithelium may show acanthosis with elongation of epidermal ridges and hyperpigmentation in the basal layer. In malignant melanoma, the brown pigment is melanin and melanophages may be present. Tumor cells are positive for melanocytic markers: HMB-45, MLANA, MART-1 and S100 [17, 18]. Changes in surface epithelium can be seen, including pagetoid migration or melanoma in situ. In Kaposi’s sarcoma, vascular proliferation may be present, often with extravasation of red blood cells and the presence of hemosiderotic pigment and hemosiderophages. The association with infection with HHV-8 and HIV, as well as increased expression of c-kit (Ki-67), represent arguments for Kaposi’s sarcoma [19, 20]. c-myc gene is correlated with vascular neoplasms and with angiogenesis, but in not correlated with Kaposi’s sarcoma [21].

The immunohistochemistry phenotype can be summarized as follows: the tumor cells express CD31, CD34, Factor VIII related antigen, podoplanin (D2-40). In case of infection with HHV-8/KSHV, latent nuclear antigen 1 can be expressed. Despite its benign nature, AFH requires regular follow-up after surgical excision and histopathological examination, as there are literature reports of local recurrences. According to Calonje and Fletcher [22], these occur in about 19% of cases. The recurrences found in AFH might be due to incomplete excision with microscopic positive margins. Estimates of the period over which local recurrence may occur vary from six weeks to 13 years [23]. The need for follow-up in these cases is due to the possibility of metastasis, as demonstrated by the same authors. Metastases have been observed up to 15 years after initial diagnosis of dermatofibroma, and occurred in the lungs, liver, lymph nodes and soft tissues [23].

Conclusions

Despite some clinical characteristics and dermoscopic features, the diagnostic criteria of aneurysmal dermatofibroma are not specific. A definite set of clinical and dermoscopically criteria for differential diagnosis for three types of tumors – aneurysmal dermatofibroma, malignant melanoma and Kaposi’s sarcoma – cannot be currently set. Wide surgical excision followed by histopathology and immunohistochemistry are required to clarify the diagnosis and exclude KS or MM. Although aneurysmal dermatofibroma is a benign skin lesion, long-term follow-up of patients is required to detect possible recurrence or metastasis.

References


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