Case Report

Multinucleate cell angiohistiocytoma versus symplastic hemangioma - diagnostic dilemma

Sankappa Pundalikappa Sinhasan, Mary Theresa Sylvia, Mima Maychet Sangma[1], Ramachandra Venkataraman Bhat

Departments of Pathology and [1]Surgery, Indira Gandhi Medical College and Research Institute, Puducherry, India

ABSTRACT

Symplastic hemangioma (SH) represents degenerative changes occurring in preexisting hemangioma that can mimic sarcomas and pseudosarcomas. Around eight cases of SH have been reported so far in the literature indicating its rare presentation. On the other hand, multinucleate cell angiohistiocytoma (MCAH) is believed to be a reactive inflammatory process rather than a true neoplastic process. Around 80 cases of MCAH have been reported so far. Both these entities share common histopathological features such as the presence of vascular channels, bizarre cells, inflammatory stroma, and multinucleate giant cells making them to masquerade sarcoma. Hereby, we report a case of a tiny swelling over the dorsum of hand in a 30-year-old female, with a clinical diagnosis of hemangioma. The histopathological examination revealed poorly circumscribed vascular lesion with bizarre cells, multinucleated giant cells scattered against the dense inflammatory background. We considered differential diagnoses of MCAH, SH, and dermatofibroma. We also considered malignancies in differential diagnoses such as hemangioendothelioma, liposarcoma, and metastatic nodule due to the presence of bizarre cells. This case report reemphasizes the presence of atypical histological features that represent degenerative changes occurring in benign tumors which can masquerade malignancies. We discuss the histological features that will help to differentiate SH and MCAH.

Key words: Bizarre cells, multinucleate cell angiohistiocytoma, symplastic hemangioma

INTRODUCTION

Multinucleate cell angiohistiocytoma (MCAH) is rare entity described in 1985 by Smith and Wilson Jones.[1] This is unusual skin condition most commonly presented by middle-aged women as small erythematous papule over limbs, mainly on the dorsum of hands. It can clinically mimic hemangiomas, Kaposi sarcoma, insect bite, lichen planus, sarcoidosis, and granuloma annulare.[2] Histopathology is challenging as it can resemble inflammatory conditions, fibrohistiocytic tumors, sarcomas, and pseudosarcomas. Differentiating MCAH from symplastic hemangioma (SH) is quite difficult, as experienced by us, due to quite similar histological features shared by both of these conditions. SH represents degenerative changes occurring in preexisting hemangioma that can mimic sarcomas and pseudosarcomas. Both these entities share common histopathological features such as the presence of vascular channels, bizarre cells, inflammatory stroma, and multinucleate giant cells making them to masquerade sarcoma.[3] Here, we discuss a skin lesion in a 30-year-old female, which was excised as a hemangioma. The case report highlights the diagnostic difficulties encountered by pathologists, although it may be a simple hemangioma for the operating surgeon. We alert presence of atypical histological features that represent degenerative changes occurring in benign tumors which can masquerade malignancies. Immunohistochemistry (IHC) is of not much useful in differentiating these two conditions. We considered few differences in these two conditions after

Address for correspondence: Dr. Sankappa Pundalikappa Sinhasan, Department of Pathology, Indira Gandhi Medical College and Research Institute, Puducherry - 605 009, India.
E-mail: drspsinhasan@gmail.com

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review of literature like thin capillary sized vessels favor MCAH whereas large cavernous sized vessels are seen in SH. Inflammation will be moderate to abundant in MCAH whereas minimal to moderate in SH. Location wise, MCAH is more common in extremities, but SH is common on scalp and face. MCAH is more often a disease of a middle-aged female, but no age specification seen for SH due to a limited number of cases reported.[4,5]

CASE REPORT

A 30-year-old woman presented to the general surgery outpatient department (OPD) with complaints of swelling over the dorsum of the hand of 6 months duration. She denied the history of trauma, itching, pain or similar lesions anywhere in the body. Local examination revealed a tiny swelling measuring 1.5 cm × 1.0 cm with reddish discoloration of the overlying skin. The swelling was soft and nontender, situated in the subcutaneous plane. Clinical diagnosis of hemangioma was made, and lesion was excised completely under local anesthesia by the surgeon on OPD basis. Patient advised for follow-up, but she did not turned-up even after 6 months after the surgery.

Grossly, the lesion was partly solid and partly cystic with gray brown areas of hemorrhages. The entire lesion was bisected and submitted for microscopic examination. Histopathological examination revealed poorly circumscribed lesion with hypo and hypercellular areas. The stroma showed myxoid degeneration and vascular channels with areas of hemorrhages on scanner view [Figure 1]. The high power showed dense inflammatory cell infiltrate composed of neutrophils, lymphocytes, eosinophils, mononuclear histiocytes and scattered bizarre appearing multinucleate giant cells [Figure 2]. Multiple deeper sections revealed cells with intranuclear vacuoles and bizarre monster cells. The myxoid areas showed cells with multi-vacuolated cytoplasm resembling "lipoblasts." Occasional cells showed eccentrically placed nucleus with intracytoplasmic mucin vacuoles resembling "signet ring adenocarcinoma cells." The cellularity, pleomorphism, inflammatory component, vascular component, myxoid degeneration varied from one area to the other. Histopathologically we considered differential diagnoses of resembled benign inflammatory condition, vascular tumor, fibrohistiocytic benign tumor, liposarcoma, hemangioendothelioma, bacillary angiomatosis, and metastatic nodule. Discussion with a clinician at this junction revealed an apparently healthy patient with no significant history. Further clinical details were not available as the case was operated on an outpatient basis. The final histopathological impression of multinucleate giant cell angiohistiocytoma was issued and asked for confirmation by IHC. A panel of IHC marker study [Figure 3] was carried out to know exact the nature of the lesion. The vimentin showed strong intracytoplasmic positivity among

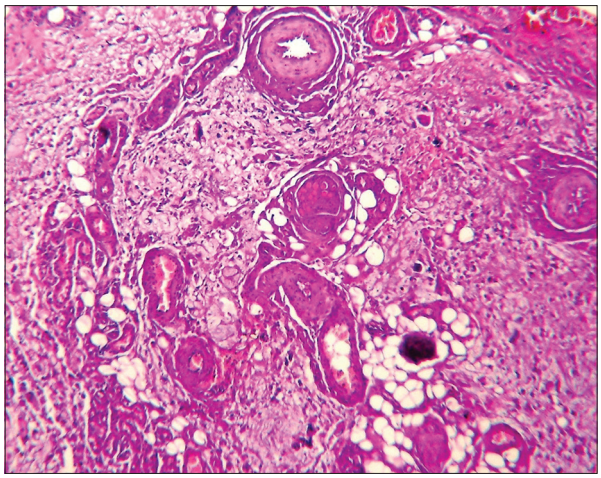

Figure 1: Poorly circumscribed lesion showing myxoid stroma containing vascular channels with areas of hemorrhages (H and E, ×4)

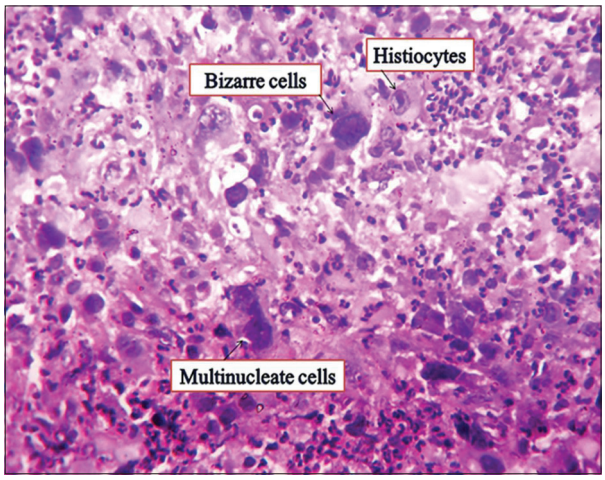

Figure 2: Bizarre appearing cells, multinucleate giant cells and histiocytes scattered against dense inflammatory background (H and E, ×40)

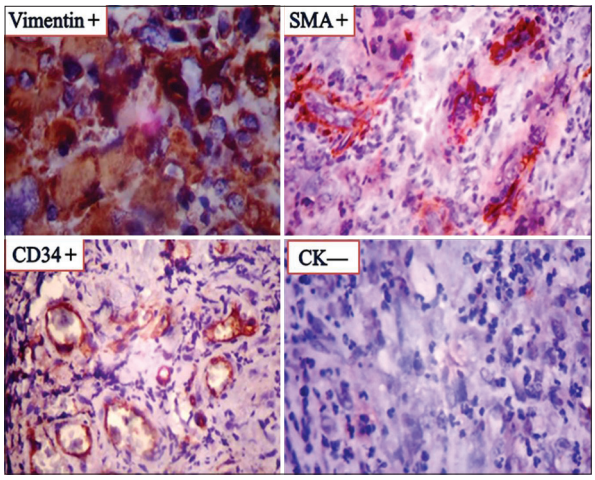

Figure 3: Immunohistochemistry showing strong vimentin positivity in stromal cells, smooth muscle actin and CD34 positivity in vascular endothelial cells and cytokeratin 7 negativity

histiocyte-like mononuclear cells as well as in multinucleate giant cells. This was indicative of fibrohistiocytic origin. Smooth muscle actin showed intense positivity in the wall of vascular channels scattered throughout the lesion. The CD34 was strongly positive for the vascular endothelial cells. These findings suggested vascular nature of the lesion. The cytokeratin 7, S-100 and Ki-67 were negative.

DISCUSSION

Around 80 cases of MCAH have been reported in the literature so far, which means that it is most likely underreported.[3] Clinically, the majority of MCAH lesions present on the extremities, but they can also appear as a painless swelling anywhere else in the body. Histopathology of MCAH shows increased number of blood vessels accompanied by inflammatory cell infiltrate, fiClinically, th mononuclear cells, and the presence of multinucleated giant cells. On the other hand, SH is a rare type of hemangioma with only eight case reports so far in the literature.[4] SH represents hemangioma with degenerative atypia. This is comparable to other benign tumors with degenerative atypia such as ancient schwannomas and symplastic leiomyomas. Similar to MCAH, multinucleate giant cells, and fibrohistiocytic cells can also be seen in SH.[6]

Histologically, MCAH can resemble a variety of soft tissue tumors due to the presence of its degenerative nuclear atypia features. The angiomatous type of Kaposi's sarcoma can also resemble MCAH because it may contain a few proliferating spindle cells. The extravasation of red blood cells (RBCs) always seen in Kaposi's sarcoma and lack of multinucleate cells helps to differentiate these two conditions. We also considered hemangioendothelioma as a differential diagnosis due to the presence of cells with intranuclear lumina resembling mucin vacuoles. However, unlike hemangioendothelioma, these lumina do not seem to contain RBC's. IHC of MCAH, the endothelial cells of the capillaries appear normal and stain positive for factor VIII related antigen, ulex europaeus I lectin, CD31, and CD34. The multinucleate cells also stain positive only with antibodies to vimentin, whereas the mononuclear interstitial cells are positive for CD68, lysozyme, and alfa-1 antitrypsin, but are negative for S-100 protein and CD1a. Some of them are positive also for monocyte-macrophage marker MAC 387. The scattered interstitial mononuclear cells, as well as the multinucleate giant cells and bizarre appearing monster cells seen in MCAH, are positive for vimentin and negative for S-100 supporting a monocyte-macrophage lineage. Probable they originate from fibroblasts and this hypothesis is supported by electron microscopic findings reported by Smolle *et al*.[5]

Ultrastructurally, the presence of a zonula nucleum limitans and a prominent rough endoplasmic reticulum in the multinucleate cells suggests fibroblastic differentiation of these cells.[5] The role of estrogen receptor α overexpression in MCAH is described by Cesinaro *et al*., who noted female predominance of the lesion and probable treatment with anti-estrogen therapy, especially for multifocal lesions.[7] Hence, estrogen receptor alpha and progesterone receptor positivity may favor the diagnosis of MCAH as both are negative in SH, but IHC alone is not of much useful in differentiating these two conditions.

Clinically, MCAH tends to be multiple, erythematous and eruptive lesions, occurring preferentially on areas subject to trauma or arthropod bites, and in some cases undergo spontaneous regression. Based on all the evidence, some investigators have suggested that MCAH is an inflammatory disorder rather than a truly neoplastic process.[7]

Surgical excision of MCAH may be recommended for cosmetic reasons. Laser therapy has resulted in clinical resolution of the lesions without scarring. Kopera *et al*. reported successful treatment with an argon laser. Treatment using intense pulsed light and cryosurgery has also been reported.[8]

CONCLUSION

MCAH has a distinctive clinicopathological feature. Histologically it mimics a variety of soft tissue tumors due to the presence of monster cells and multinucleate giant cells against myxoid stroma. However, careful evaluation of lesion will reveal its benign vascular nature with nuclear atypia due to degenerative changes. SH also shares many histological features of MCAH, but it shows the presence of cavernous sized vessels and preferences to appear on the scalp. The IHC markers may not be very useful in differentiating these two entities but significantly helps to roll-out variety of soft tissue mimickers.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Smith NP, Wilson Jones E. Multinucleate cell angiohistiocytoma: A new entity. Br J Dermatol 1985;113:15.

2. Väkevä L, Saksela O, Kariniemi AL. Multinucleate cell angiohistiocytoma: A report of four cases in Finland. Acta Derm Venereol 2003;83:222-3.
3. Wang CP, Su HY, Wu YH, Lin YC. Multinucleate cell angiohistiocytoma: A case report. Dermatol Sin 2005;23:154-7.
4. Kamarashev J, French LE, Dummer R, Kerl K. Symplastic glomus tumor – A rare but distinct benign histological variant with analogy to other 'ancient' benign skin neoplasms. J Cutan Pathol 2009;36:1099-102.
5. Smolle J, Auboeck L, Gogg-Retzer I, Soyer HP, Kerl H. Multinucleate cell angiohistiocytoma: A clinicopathological, immunohistochemical and ultrastructural study. Br J Dermatol 1989;121:113-21.
6. Goh SG, Dayrit JF, Calonje E. Symplastic hemangioma: Report of two cases. J Cutan Pathol 2006;33:735-40.
7. Cesinaro AM, Roncati L, Maiorana A. Estrogen receptor alpha overexpression in multinucleate cell angiohistiocytoma: New insights into the pathogenesis of a reactive process. Am J Dermatopathol 2010;32:655-9.
8. Kopera D, Smolle J, Kerl H. Multinucleate cell angiohistiocytoma: Treatment with argon laser. Br J Dermatol 1995;133:308-10.