Multinucleate Cell Angiohistiocytoma: A Case Presentation and Discussion

Angela Macri, DO,* Jaclyn Hess, MD,** Jonathan S. Crane, DO, FAOCD, FAAD

*Dermatology resident, PGY-2, OMNEE / Sampson Regional Medical Center, Clinton, NC
**Rotating dermatology intern, PGY-1, New Hannover Regional Medical Center, Wilmington, NC
***Program director, Dermatology Residency Program, OMNEE / Sampson Regional Medical Center, Clinton, NC; Assistant professor, Dermatology, Campbell University School of Osteopathic Medicine, Buies Creek, NC; Senior attending, New Hanover Regional Medical Center, Wilmington, NC

Disclosures: None
Correspondence: Angela Macri, DO; angmacri17@gmail.com

Abstract

Multinucleate cell angiohistiocytoma is a rare, benign disease that occurs in middle-aged women and presents as reddish-brown to violaceous, dome-shaped papules on the extremities or face. These lesions can progress, and may become disseminated. Several therapies have been documented in the literature, with varying results. We present this case to increase awareness of multinucleate cell angiohistiocytoma and show that the KTP laser is an effective treatment for this rare condition.

Introduction

Multinucleate cell angiohistiocytoma is a rare, benign disease in which asymptomatic, reddish-brown to violaceous, dome-shaped papules develop over weeks to months, commonly on the extremities and face. Lesions can be distributed in a random, linear, or annular pattern1-3 and can rarely present generalized.4 To date there are fewer than 150 reported cases of this in international literature.6 This condition is found most frequently in middle-aged to elderly women, with a median age of 55 years old.6 There are no definitive treatments, but a number of different therapies have been attempted. Previously documented therapies include vascular lasers and topical steroids, although no one treatment has been shown to be superior.

Presentation

A 69-year-old white female with a history of hypertension presented with slow-growing lesions on her left medial knee that were tender, itchy, and progressing (Figures 1 and 2). They began appearing four years prior. Biopsies were obtained and demonstrated a proliferation of small, thin-walled vascular channels, surrounded by pericytes in the superficial and mid dermis (Figure 3). The surrounding dermis contained scattered multinucleated cells with angulated cytoplasm (Figure 4) and a background of somewhat hyalinized collagen bundles. A few inflammatory cells were noted. An immunostain for CD31 was used to highlight the vascular proliferation. HHV8 immunostain was negative. The findings were consistent with multinucleate cell angiohistiocytoma. A potassium titanyl phosphate (KTP) laser with a wavelength of 532 nm was used to treat the lesions, with a 2 mm spot size, fluence of 10 J/cm2, a repetition rate of 5 Hz, and a 10 millisecond pulse duration. Two separate treatment sessions were completed one month apart (Figures 5 and 6).

Discussion

Multinucleate cell angiohistiocytoma was first described in 1985 by Smith and Wilson Jones.7 In 2015, a review of 142 cases by Frew found that 79% of patients were female, and the most commonly affected areas were the hands (30%), face (29%), legs (20%) and abdomen (10%). Clinically, these lesions may resemble Kaposi’s sarcoma, acroangiodermatitis, granuloma

![Figure 1](image1)
![Figure 2](image2)
![Figure 3](image3)
![Figure 4](image4)
![Figure 5](image5)
![Figure 6](image6)
MCA is considered to be of fibrohistiocytic origin, and there is considerable histological overlap with entities such as fibrous papule of the nose, the vascular atrophic variant of dermatofibroma, and others. The main histologic findings include bizarre basophilic multinucleated cells, small-vessel inflammation, mild dermal fibrosis, epidermal hyperplasia, and sparse lymphohistiocytic infiltrate. Immunohistochemical studies are variable in their results but have shown that vascular endothelial cells stain positive for CD68, normally a macrophage or histiocytic marker, as well as for factor VIII, CD31, and CD34. The multinucleated cells are negative for endothelial markers factor VIII and CD34 and positive for macrophage/histiocytic markers factor XIIa and CD68.

The pathogenesis of MCA may be an underlying inflammatory stimulus that leads to vascular changes and eventually a fibrotic response that precipitates further lesion development. CD68 positivity in endothelial cells may be related to an inflammatory response involving migrating macrophages that are responsible for the increased dermal vascularity. Findings of macrophages and fibrosis are consistent with lesions that are longstanding, which may be why some multinucleated cells in MCA are negative for CD68. An overexpression of estrogen receptor alpha has been found in spindle cells and multinucleated cells of MCA, also suspected to be responsible for the vascular hyperplasia. The overexpression of estrogen receptors in these lesions may be why MCAs are found predominantly in females.

MCA slowly progresses, but some cases have spontaneously resolved. There has been no statistically significant association between MCA and neoplasia. Several treatments have been used for MCA, including argon laser, intense pulsed light, carbon dioxide laser, cryosurgery, surgical excision, and intralesional steroids in combination with pulsed dye laser. Because of the progressive nature and disturbing cosmetic appearance of the lesions, our patient requested treatment. We used the 532 nm potassium titanyl phosphate laser; to our knowledge, this is the first documented case of this laser being used to treat MCA. It was chosen based on its ability to treat superficial vascular lesions, as vascular proliferation is a prominent finding in MCA. The patient underwent two treatments with the KTP laser and reported a decrease in size and erythema and a smoother texture to the lesions. She experienced only slight pain at the time of the procedure but did not experience any bruising, a benefit of the KTP compared to the PDL laser. She underwent two treatments and experienced approximately a 50% overall improvement in the appearance of the lesions. We suspect that with further treatment she will improve even more.

**Conclusion**

Multinucleate cell angiohistiocytoma is a rare, benign condition that may be under-recognized by dermatologists. Because of its progressive nature and disturbing cosmetic appearance, treatment of these lesions is often desired. Our case shows that the KTP laser is a practical treatment option for MCA based on its ability to treat superficial vascular lesions.

**References**