Letters to the Editor

ostia in the lesional areas which is in keeping with the mosaic nature of the disorder.[3]

In conclusion, our case emphasizes that linear and whorled nevoid hypermelanosis may be dermoscopically heterogeneous, displaying some individual variations. It also confirms that dermoscopy might be useful in differentiating this disorder from its prime differential diagnosis, namely, incontinentia pigmenti, which is characterized by blue-gray dots histologically corresponding to pigmentary incontinence.[3]

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Multifocal spindle cell hemangioma: Report of two cases

Sir,
Spindle cell hemangioma, formerly known as spindle cell hemangioendothelioma was first described in 1986 and was initially considered to be a low-grade angiosarcoma. In 1996, its benign behavior was recognised and the name spindle cell hemangioma was proposed.[1] It is a rare vascular proliferation, with less than 200 reported cases. The tumor originates from blood vessels and recently, lymphatic origin has been suggested.[2] The standard treatment is wide local excision but the the tumor has a high recurrence rate. We present two cases of multifocal spindle cell hemangiomas. One case was successfully treated with sclerotherapy suggesting that this can be an alternative treatment which is less invasive and has better long-term results.

The first case was a 10-year-old girl with a 4-year history of multiple violaceous and reddish cutaneous and subcutaneous nodules on the right ankle and foot [Figure 1]. Magnetic resonance imaging performed to determine the extent of the lesions showed multiple hypervascular deep nodules in the left foot and ankle with irregular bone resorption. She was treated with sclerotherapy. The nodules were punctured under ultrasound guidance. Under angiographic monitoring, injection of 2% etoxisclerol foam was performed [Figure 2] which lead to significant reduction in the volume of the lesions. There were no adverse effects and patient was asymptomatic at 2 years of follow up.

The second case was an 18-year-old woman with multiple subcutaneous nodules located on the left upper extremity. Magnetic resonance imaging showed multiple hyperintense nodules on T2 in the left upper extremity and neck. Patient had undergone surgical removal of 12 nodules 4 years back, all of which had recurred in 18 months and new lesions continued to appear.

In both cases, no enchondromas were found in the affected bones ruling out Maffucci’s syndrome.

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The first case was a 10-year-old girl with a 4-year history of multiple violaceous and reddish cutaneous and subcutaneous nodules on the right ankle and foot [Figure 1]. Magnetic resonance imaging performed to determine the extent of the lesions showed multiple hypervascular deep nodules in the left foot and ankle with irregular bone resorption. She was treated with sclerotherapy. The nodules were punctured under ultrasound guidance. Under angiographic monitoring, injection of 2% etoxisclerol foam was performed [Figure 2] which lead to significant reduction in the volume of the lesions. There were no adverse effects and patient was asymptomatic at 2 years of follow up.

The second case was an 18-year-old woman with multiple subcutaneous nodules located on the left upper extremity. Magnetic resonance imaging showed multiple hyperintense nodules on T2 in the left upper extremity and neck. Patient had undergone surgical removal of 12 nodules 4 years back, all of which had recurred in 18 months and new lesions continued to appear.

In both cases, no enchondromas were found in the affected bones ruling out Maffucci’s syndrome.
The histopathological findings in both patients showed nodular proliferation of thin-walled vascular structures lined by a single layer of endothelial cells with small nuclei of uniform appearance with some vessels showing dilation. Between the vascular spaces, fascicles of monomorphic spindle cells without nuclear atypia were present; these stained negatively for CD31 and positively for actin. Clusters of round cells with vacuolated cytoplasm positive for the CD31 were also seen [Figure 3]. Immunohistochemistry for human herpes virus type 8 was negative ruling out Kaposi’s sarcoma.

Spindle cell hemangioma is a rare, benign, vascular lesion which is more common in children and young adults, with no sex predilection. It presents mostly in the dermis and subcutaneous tissue of distal extremities. It has also been reported intramuscularly and exceptionally, it can affect the viscera such as the pancreas. Clinically it presents as a single, or sometimes multiple nodules that can be painful. These nodules are small, varying in size from 0.5 to 2.5 cm in diameter, skin colored or bluish, soft, firm in consistency and slow-growing. Multiple lesions have been associated with Maffucci’s syndrome, a sporadically inherited genetic disorder where multiple spindle cell hemangiomas can be associated with phleboliths and multiple enchondromas.[3]

The histopathological findings show relatively well-circumscribed but non-encapsulated, dermal or subcutaneous nodules composed of thin-walled dilated vessels lined by flat endothelial cells, sometimes with thrombi or phleboliths in their lumina.[4] These areas alternate with areas of monomorphic spindle cells which are arranged in fascicles and have uniform, elongated, dark nuclei with eosinophilic cytoplasm. They do not show cytological atypia and mitotic figures are rarely found. There can be intravascular involvement. Variable numbers of epitheloid cells can also be present.

Immunohistochemistry shows that the cells lining the vascular spaces are positive for factor VIII-related antigen, CD31 and the hematopoietic progenitor cell antigen (CD34) markers. On the other hand, the spindle cells are negative for these markers and can be positive for actin supporting the mesenchymal origin of these cells.[4]

A recent study has shown the expression of homeobox transcription factor (Prox-1) and lymphatic endothelial cell marker podoplanin (D2-40) in these lesions, suggesting that these are proliferations of lymphatic origin.[2]

In the differential diagnosis, other vascular tumors should be considered such as epitheloid hemangiendothelioma, Kaposi’s sarcoma and spindle cell angiosarcoma, among others.

The clinical course is typically indolent but in spite of its benign behavior, the development of new lesions in the same anatomical area after surgical treatment is >50%.[1] This phenomenon is considered by some authors not as recurrences but as new primaries.[1,5] Successful treatment has also been reported with post-operative radiotherapy, low-dose intralesional interferon α-2b and intra-arterial administration of recombinant interleukin-2.[3]
It is important to recognize spindle cell hemangioma because aggressive treatment can be avoided. Even though the standard treatment is wide local excision, due to the high rate of recurrences, we propose sclerotherapy as an alternative management strategy that may provide satisfactory results.

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