Letters to the Editor

Observation Letters

Linear basal cell carcinoma: Report of three cases with dermoscopic findings

Sir,

Linear basal cell carcinoma was first described as a new clinical subtype in 1985 by Lewis. We were able to find published reports of only 43 other cases since then. Certain unique clinical and histological characteristics differentiate it from other basal cell carcinomas hence, some authors consider it as a different clinical entity.[1,2] Herein, we report three cases of linear basal cell carcinoma.

Patient 1 was an 86-year-old woman referred for the evaluation of a linear pigmented lesion on the right side of the neck. The lesion was first noticed 10 months previously; there was no evidence of trauma or any other trigger at that site. On physical examination, we observed a linear pigmented macule aligned along the relaxed skin tension lines, measuring 10 mm × 1 mm [Figure 1a]. Dermoscopy highlighted the absence of a pigmented network pattern and the presence of three of the six criteria for pigmented basal cell carcinoma according to Menzies: maple leaf-like areas, spoke-wheel areas and large blue-gray ovoid nests [Figure 1b].[3] Patient 2 was an 84-year-old man who consulted us for an asymptomatic, 16 mm × 4 mm lesion on his upper chest which had slowly enlarged in a linear fashion in the last 2 years [Figure 2a]. Dermoscopy showed maple leaf-like areas, spoke-wheel areas, large blue-gray ovoid nests, ulceration and arborizing telangiectasias [Figure 2b]. Patient 3 was a 77-year-old woman who presented with a 13-month history of an asymptomatic lesion on her face. We observed a non-palpable, linear pigmented lesion measuring 14 mm × 2 mm [Figure 3a]. Dermoscopic findings included maple leaf-like areas, spoke-wheel areas, large blue-gray ovoid nests and slight ulceration [Figure 3b]. In our three patients, the tumor region was completely excised with a 3-mm safety margin with primary closure of the wound. The histological findings confirmed the diagnosis of basal cell carcinoma; the surgical margins were free of tumor cells in all cases. Histologically, in patient 1 and 2, the tumour nests mainly appeared in a nodular arrangement [Figures 1c and 2c], so nodular subtype of basal cell carcinoma was diagnosed. In patient 3, the histological analysis of the lesion was diagnostic of the superficial variant of basal cell carcinoma [Figure 3c].

Linear basal cell carcinoma extends preferentially in one direction resulting in the characteristic morphology of a linear lesion with straight edges and a length-to-width ratio of at least 3:1.[4] Some of the mechanisms that have been suggested to explain this mode of growth include limitation of lateral spread of the lesion by dermal fibrosis or interaction of the stroma with Langer’s lines since basal cell carcinomas depend on stromal interactions...
for progression and growth. Focal trauma is an established risk factor in the development of basal cell carcinoma so the possibility of Koebnerisation has also been proposed to explain the linear pattern and its orientation along the tension lines of the skin.\cite{1,2} In the largest histological series, the proportion of basal cell carcinoma containing pigment ranges from 6.7% to 8.5%.\cite{5} In our cases, 100% of the tumors were pigmented and showed maple leaf-like areas and spoke wheel areas on dermoscopy, two uncommon but highly specific diagnostic criteria. Among the reported cases, the most common histological subtype was nodular.\cite{2} However, Al-Niaimi and Lyon found that 32% of linear basal cell carcinoma had an aggressive histological subtype such as micronodular, infiltrative or morpheaform; a percentage much greater than expected in the general population.\cite{1} In addition, some authors have described the potential for wider subclinical extension in this variant of basal cell carcinoma. Therefore, Mohs micrographic surgery is preferred as the treatment of choice.\cite{2} In our three patients, the histological findings did correlate with the clinical appearance of the lesions.
and conventional surgery was curative. We think this clinical subtype of basal cell carcinoma should not determine the surgical approach. Conventional surgery can be a suitable first-line treatment and Mohs micrographic surgery should be reserved as an alternative in the following scenarios: anatomical regions where large resection margins are not practicable, recurrent forms of basal cell carcinoma, when unexpected positive margins occur after recent excision, primary aggressive histological forms of the tumor: morpheaform, infiltrating, perineural, metatypical or micronodular, or high-risk areas of body: area H and area M. We believe this entity is under-reported due to its under-recognition. Further investigation of linear basal cell carcinoma in a larger series of patients is needed to elucidate the histological aggressiveness of this clinical subtype, the proportion of linear basal cell carcinomas containing pigment and the best surgical approach for these patients. Undoubtedly, dermoscopy is an essential tool in the diagnosis of this entity.

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Conflicts of interest
There are no conflicts of interest.

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Sir,

Waardenburg syndrome is a rare genetic disorder of neural crest cell development characterized by the association of craniofacial dysmorphisms (synophrys, telecanthus, broad and high nasal root and lower lacrimal dystopia), pigmentation defects (heterochromic and bright hypochromic blue irides, hypopigmented spots on the skin and white forelock or early graying) and sensorineural congenital hearing impairment.

Four variants of Waardenburg syndrome have been described. Of these, Waardenburg syndrome 1 and 2 are the most common variants and are inherited in an autosomal dominant pattern in most of the cases.

Leukoderma (hypopigmented patches) is a characteristic feature of Waardenburg syndrome and hence is included in the widely accepted diagnostic criteria proposed by the Waardenburg consortium [Table 1].

We report a case of Waardenburg syndrome 1, having a predominance of multiple hyperpigmented patches all over the body along with only a few hypopigmented patches.

A 9-month-old boy, the first child of a non-consanguineous marriage, was born following a full-term uncomplicated vaginal delivery and presented with a white forelock and multiple brown-colored areas all over the body since birth. There were two first-trimester abortions in the mother prior to this pregnancy. Family history was not contributory. The child was active and feeding well.

Clinical examination revealed a white forelock (poliosis) in the frontal area with an underlying depigmented patch [Figure 1a]. There were multiple, ill-defined, dark brown, hyperpigmented patches ranging from 0.5 cm to 7 cm all over the body, relatively sparing the face [Figure 2]. In addition, a few ill-defined hypopigmented patches with islands of hyperpigmentation within them were noted on the lower abdomen and thighs [Figure 3]. The nasal root was broad. On ophthalmological examination, dystopia canthorum and bilaterally blue iridis were found [Figure 1b]. Systemic examination including cardiovascular and orthopedic examination was normal and routine laboratory investigations including complete blood count, liver and kidney function tests, chest X-ray, electrocardiogram and echocardiography were normal. Brainstem evoked response audiometry revealed bilateral sensorineural hearing loss. Physical development and milestones were appropriate for age.

The diagnosis of Waardenburg syndrome 1 was made as per the Waardenburg Consortium criteria. Owing to the presence of multiple hyperpigmented patches, conditions such as LEOPARD syndrome and neurofibromatoses were considered as differential diagnoses but were subsequently ruled out on...

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