Case Report

Seborrhoeic keratosis with autosomal dominant inheritance - a rare case report

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ABSTRACT

Seborrhoeic keratosis is a benign tumour composed of epidermal keratinocytes, displaying varying morphological features, frequently pigmented and more common in the elderly. Various clinical and histopathological variants have been described. A genetically determined predisposition to Seborrhoeic keratosis is largely accepted. We report a case of multiple Seborrhoeic keratoses of childhood onset and transmitted for three generations in her family members as an autosomal dominant trait.

Keywords: Autosomal dominant inheritance, Childhood onset, Multiple seborrhoeic keratoses

INTRODUCTION

Seborrhoeic keratosis is a benign non-melanocytic epidermal tumour composed of keratinocytes. Seborrhoeic keratosis is seen more commonly in white skinned people, usually appears in the fifth decade with equal sex preponderance.1,2 The disease displays varying morphological features and histopathological types. Family history should be sought if there are multiple seborrhoeic keratoses, and if associated with an early age of onset.3 Here we report a case of multiple seborrhoeic keratoses in a young female, in whom the disease started at a very early age and similar complaints in some of her family members.

CASE REPORT

A 17-year-old unmarried female came to our outpatient department with H/O multiple dark colored raised skin lesions over the face, neck, trunk and extremities since the age of 7 years. Patient has been apparently normal till 7 years of age after which she had developed a single dark coloured raised skin lesion over front of chest. Later she had developed multiple similar lesions of varying sizes over trunk, face, neck and extremities. Few lesions had gradually increased in size over the past 10 years.

Figure 1: Multiple, well defined, raised, hyperpigmented papules and plaques of varying sizes, with rough surface and stuck on appearance seen over left side of face.
There was no H/O itching, pain or photosensitivity over the lesions. There was no H/O sudden change in size, morphology or ulceration seen in any of the lesion over the years. Patient also gave H/O of her maternal grandmother, mother and maternal uncle also had similar skin lesions from their childhood. On examination patient’s vitals were stable. General and systemic examination were normal.

Examination of other family members of the patient also revealed similar findings (Figure 2-4). Histopathological examination of the lesional skin showed hyperkeratosis, acanthosis, papillomatosis, melanocytic proliferation and pseudo horn cyst, consistent with acanthotic variety of seborrhoeic keratosis (Figure 5).

Figure 2: Multiple Seborrhoeic keratosis over left side of face and forehead (Patient’s mother).

Figure 3: Multiple seborrhoeic keratosis over left temporal region and left side of forehead (Patient’s maternal uncle).

On dermatological examination, multiple, well defined, raised, hyperpigmented papules and plaques of varying sizes, with rough surface and stuck on appearance seen all over the body (Figure 1). Her scalp, hair, nails, palms and soles and mucosae were normal.

 Examination of other family members of the patient also revealed similar findings (Figure 2-4). Histopathological examination of the lesional skin showed hyperkeratosis, acanthosis, papillomatosis, melanocytic proliferation and pseudo horn cyst, consistent with acanthotic variety of seborrhoeic keratosis (Figure 5).

HPE of the skin lesions of other family members were also suggestive of seborrhoeic keratosis. Mode of transmission of the disease among the family members has been depicted in the pedigree chart (Figure 6).

Figure 6: Pedigree chart showing the mode of inheritance of the disease among the family members.

USG abdomen was taken for all the affected family members which didn’t show any evidence of internal malignancy. There was no history or examination findings suggestive of any other inflammatory dermatoses in the patient and her family members. Patient and her family members have been explained about the disease, reassurance given and has been advised regular follow up.

DISCUSSION

Seborrhoeic keratosis is a benign non-melanocytic epidermal tumour composed of keratinocytes, displaying
varying morphological features, frequently pigmented and more common in the elderly. Seborrhoeic wart, Senile wart, Basal cell papilloma, Senile keratoses, Seborrhoeic verruca, Barnacles of old age are some of the other names for seborrhoeic keratosis.

Seborrhoeic keratosis (SKs) is seen more commonly in white skinned people. It usually appears in the fifth decade, but also seen in middle age in tropical countries with equal sex preponderance. The total number and size of seborrhoeic keratosis increases with increase in age. Family history of SKs should be sought if there are multiple SKs and if associated with an early age of onset. Sunlight and Human Papilloma Virus (HPV) has been implicated in the causation of seborrhoeic keratosis. Genetic predisposition is seen and the disease is inherited in families as an autosomal dominant trait. Activating mutations in FGFR3 which provide proliferative signals to keratinocytes, increased expression of Bcl2 and Ki-67, increased DNp63a expression and activating PIK3CA mutations has been noted in the pathogenesis of SKs. Increased expression of keratinocyte derived Endothelin I mediated by TNF α and Endothelin Converting Enzyme 1α (ECE 1α) is linked to pigmentation seen in SKs.

Seborrhoeic keratoses are usually asymptomatic, sometimes pruritic, starts as well defined round to oval slightly hyperpigmented papules or plaques ranging in size from few millimeters, but rarely exceeding 3 cm. It is commonly seen over face and trunk but also seen in neck and extremities. Nipple, areola, vulva and conjunctiva were other unusual sites where SKs were seen. Mucous membranes are spared in SKs. Unusual distribution patterns seen with SKs are lesions along skin cleavage lines over the back and waist, dermalomal distribution, streamline arrangement, naeviform fashion and also as rain drop like lesions crossing the skin cleavage lines.[3] In a study from South India, Common SK (60%), DPN (46.4%), Pedunculated (21.2), Flat (10.4%) and Stucco keratosis (2%) were the clinical variants seen. The surface of these SKs has numerous plugged follicular orifices giving an almost cerebriform appearance. They vary from dirty yellow to black in colour and may have a typical “stuck on appearance”. They do not reflect light differing from naevi.

Lentigo maligna, actinic keratosis, melanocytic naevi, malignant melanoma and pigmented BCC are some of the differential diagnosis for SKs. Seborrhoeic keratoses are usually diagnosed clinically. By dermatoscopy various patterns were seen, which are helpful in differentiating SKs from other pigmented tumours. Comedo like openings (CL), Fissures and ridges (FR) and Sharp demarcation (SD) were consistent findings on dermatoscopy in common SKs. Other findings were Finger print (FP), Moth eaten borders (ME), milia like cysts (ML) and network like structures (NL). DPN shows only three patterns CL, FR and SD. Pedunculated SKs shows only two patterns FR and CL. Stuccokeratosis shows SD and NL in all cases. Skin biopsy shows hyperkeratosis, acanthosis with immature keratinocytes, papillomatosis with church steeple appearance, melanocytic proliferation, pseudo horn cysts with lower border which lies in a straight line with normal epidermis. Six histopathologic variants are seen – Hyperkeratotic, Acanthotic, Adenoid or Reticulate, Clonal, Irritated and Melanoacanthoma.

Infection, bleeding, oozing and crusting are some of the complications associated with SKs. Cascoa et al found superficial BCC, SCC and Malignant melanoma in situ associated with few cases of seborrhoeic keratosis. Sudden appearance of multiple eruptive SKs “Lesser – Tretl sign” is associated with Gastric adenocarcinoma, due to the growth factors secreted by the tumour. Treatment options include curettage, cryotherapy, ablative and non-ablative lasers, dermabrasion, 5 FU and medium depth peels. Treatment sought mainly for cosmetic reasons. Dyspigmentation and recurrence may occur following lesional clearance.

CONCLUSION

Though seborrhoeic keratoses are the most common benign epidermal tumours and their genetic predisposition is largely accepted, their familial occurrence has been rarely reported in literature. We present this case because of its rare combination of occurring in families along with a very early onset (childhood onset) in all the affected family members.

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REFERENCES
