Bracketed Benevolent, Bleb-Milia

Anubha Bajaji*

Histopathologist in A.B. Diagnostics, New Delhi, India

*Corresponding Author: Anubha Bajaji, Histopathologist in A.B. Diagnostics, New Delhi, India.

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Abstract

Multiple, miniature, benign, superficial, keratin impacted, epidermoid cysts derived from pilosebaceous apparatus or eccrine sweat ducts are cogitated as milia. Hypothesis for milia generation incorporate physical deterioration and microscopic inflammation of skin induced by viral (herpes zoster) or bullous disorders. Milia are classified as “primary” with spontaneous lesions of unknown origin, “secondary” when milia occur sequential to cutaneous trauma or associated injuries”, milia en plaque” where primary milia are represented as localized plaques and multiple eruptive milia (MEM) with characteristic, abrupt eruption of innumerable milia.

Keywords: Multiple Eruptive Milia (MEM); Herpes Zoster

Preface

Milia are described as multiple, miniature, benign, superficial, keratin impacted epidermoid cysts with a magnitude of one millimetre to 2 millimetres. Milia are contemplated as a derivative of pilosebaceous apparatus or eccrine sweat ducts.

Disease characteristics

Milia as a benign condition demonstrates an equivalent gender incidence. Lesions can appear at any age and are usually devoid of racial predilection.

Contingent theories for genesis of milia incorporate factors such as physical deterioration and microscopic inflammation of skin induced by viral (herpes zoster) or bullous disorders. Aforesaid pathologies contribute to the appearance of milia and epidermal cysts.

Administration of immune suppressive drugs can engender milia and epidermal cysts by enhancing inflammatory egress along with occlusion of the pilosebaceous unit and an imbalance of aforesaid factors [1,2].

"Wolf’s isotopic response" is a phenomenon describing the emergence of an independent disorder at the site of previous, alleviated cutaneous disease, frequently a herpes virus infection [3].

Clinical elucidation

Milia comprise of multiple, superficial keratinous cysts appearing as yellowish, whitish or skin coloured, dome shaped nodules or firm, discreet, disseminated or confluent papules with a centroidal occlusion and a magnitude of one millimetre to 3 millimetres. Lesions are frequently cogitated on the forehead, glabella, cheeks, vulva, scrotum or penile shaft. Lesions are essentially asymptomatic or occasionally inflamed and appear abruptly within a few weeks [3,4].

Milia are classified as:

- Primary milia where the lesions arise spontaneously and are of unknown origin. Primary milia are cogitated in an estimated 50% neonates, arise on the face, upper trunk and extremities and display a spontaneous regression. Incriminated older children and adults demonstrate apparent lesions on the face, particularly forehead, eyelids, cheeks and external genitalia. Persistent infantile milia can be associated with steatocystoma multiplex and eruptive vellus hair cyst.

- Secondary milia occur sequential to trauma to the skin or associated injuries. Secondary milia arise as a complication to several conditions such as follicular mucinosis, folliculotropic mycosis fungoides, lichen sclerosus, radiotherapy, herpes zoster infection, leishmaniasis, severe burns, dermabrasion, chemical peeling, cutaneous local steroid therapy (clobetasol propionate), systemic medication (cyclosporine), adverse drug reaction (benaxaprofen) and contact dermatitis.

Milia can emerge contingent to sub-epidermal disorders with blisters such as dystrophic epidermolysis bullosa, epidermolysis bullosa acquisita, porphyria cutanea tarda and pseudoporphyria.

Additionally, milia appear as component of familial dermatosis such as Rombo’s syndrome (facial anetoderma vermiculatum, telangiectasia, milia, hypotrichosis, acral erythema, cyanosis and a predisposition to trichoepithelioma and basal cell carcinoma), Bazex-Dupre-Christol syndrome (follicular atrophoderma, congenital hypotrichosis, basal cell carcinoma) along with conditions such as familial multiple cylindromas, trichoepithelioma, milia and spiradenomas [2,4].

Milia en plaque

Exceptionally, primary milia are represented as localized plaques. An oedematous, erythematous plaque is cogitated with an epithelial infiltration within the base and a superimposition with innumerable milia.

Lesions of the particular variant are frequent on periauricular and periorbital regions or the supraclavicular region, especially in adults. Milia en plaque can appear concomitant to pseudoxanthoma elasticum or lupus erythematosus. Essentially of obscure aetiology, milia en plaque can be incited with spectacles, earrings or perfume.

Eruptive milia are an infrequent variant, especially in children. A contemporary classification with familial incidence and an autosomal dominant mode of disease transmission or a sporadic emergence are cogitated. Eruptive milia also appear as a component of genodermatosis.

Multiple eruptive Milia (MEM) is a condition with a characteristic, abrupt eruption of innumerable milia, generally emerging within weeks or months. Aforesaid milia are exceptional, arise spontaneously and are termed as idiopathic although can be a familial variant of autosomal dominant inheritance or appear as a component of genodermatosis.

Idiopathic (spontaneous) multiple eruptive milia is an exceedingly infrequent disorder appearing in an age range of 9 years to 91 years. An equivalent gender distribution is exemplified, akin to adjunctive lesions. Lesions can additionally emerge on the scalp, neck, upper chest, back, arms, shoulders, abdomen and axillae [4,5].

Idiopathic multiple eruptive milia is a disease of obscure aetiology.

Susceptibility to sunlight can induce lesions on face, neck or shoulders. Scratching and rubbing are incident in lesions of protected zone of the body. Familial instances of multiple eruptive milia are an autosomal dominant mode of disease transmission.

Multiple eruptive milia is concomitant with genodermatosis such as Rombo’s syndrome, basal cell nevus syndrome, Bazex-Dupre-Christol syndrome, pachyonychia congenita and Gardner’s syndrome [5,6].

Histological elucidation

Milia comprise of miniature epidermoid cysts commonly situated within the superficial dermis with superimposed, integrated stratified squamous epithelium.

Multiple cysts are cogitated which are layered with stratified squamous epithelium, are devoid of adnexal structures and are impacted with eosinophilic, loosely configured, concentric, keratinous lamellae which are a typical histological feature of milia. Additionally, ruptured epidermal cysts layered with stratified squamous epithelium and an intact, granular cell layer are enunciated.

Neonatal milia frequently depict an adherence to vellus hair follicles. Secondary lesions appear in conjunction with hair follicles or an eccrine sweat duct. In fact, milia accompanied by scarring or blister associated disorders demonstrate a concordant presence of eccrine sweat ducts.

Lesions of milia en plaque classically exhibit an intermingled, intense T cell lymphocytic infiltrate [6,7].

Pathogenic associations

With Wolf’s isotopic response, multiple epidermal cysts and milia arise and are superimposed upon sites of previously recovered herpes zoster infection. Healthy individuals are implicated. Subjects demonstrate a cluster of pruritic, erythematous papules predominantly on the extremities, although no site is exempt. Viral infection incriminates cutaneous nerve fibres, nerve endings and dispersion of Langerhans cells, thereby consequently modifying the localized immune environment.

Allogeneic renal transplant with subsequent immune-suppression can frequently delineate isotopic responses with the emergence of granuloma annulare and adjunctive granulomatous responses. Additionally, comedonic- microcystic reactions and epidermal cysts can occur. Milia or multiple epidermal cysts can arise following the discernment of viral incrimination with herpes zoster [5,7].

Preceding, painful, vesiculopapular eruption initiated by herpes zoster infection is discovered at identical sites. Deteriorated skin due to a viral infraction or preceding disease is implicated with the appearance of secondary, independent disorder as the implicated area depicts an immunologic vulnerability.

Multiple, unilateral, erythematous or skin toned, miniature papules display a linear form of dissemination. The papules exhibit an inky, centralised orifice.

Wolf’s post herpetic isotopic response aggravates the generation of milia and epidermal cysts. With augmented life expectancy and immune suppression incidence of herpes zoster is elevated with consequent amplification of Wolf’s post herpetic isotopic response and emergence of milia and multiple epidermal cysts [8,9].

Genodermatosis associated with milia

- Bazex-Dupre-Christol syndrome
- Rombo syndrome
- Brooke Spiegler syndrome
- Orofaciodigital syndrome type I
- Atrichia with papular lesions
- Hereditary vitamin D- dependent rickets type IIA
- Pachyonychia congenita type II
- Basal cell nevus syndrome

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- Generalized basaloid follicular hamartoma syndrome
- Familial milia and absent dermatoglyphics
- Nicolau-Balus syndrome
- Hypotrichosis with light coloured hair and facial milia
- KID syndrome
- Epidermolysis bullosa
- Hereditary porphyrias [8].

**Differential diagnosis**

Multiple eruptive milia necessitate a segregation from clinical disorders such as eruptive syringoma, miliaria crystallina, eruptive vellus hair cyst and generalized milia like idiopathic calcinosis cutis [2,4].

**Therapeutic options**

Intense pruritus and inflammation can be adequately managed with intra lesional administration of triamcinolone. Topical application of 0.05% tretinoin cream demonstrates an appreciable outcome. For asymptomatic lesions of milia treatment is generally not indicated.

Therapeutic options applicable for milia such as exudation of lesion contents, curettage, electrodessication and cryotherapy are contemplated as unsatisfactory for multiple eruptive milia as numerous lesions necessitating treatment may not be comprehensively curative and can induce uneasiness within the individual [9,10].

Topical application of the tretinoin cream appears to be beneficial. Symptomatic epidermal cysts can be surgically excised and milia can be appropriately cauterized.

Milia en plaque is suitably managed with oral etretinate which is also beneficial in multiple eruptive milia.

Oral azithromycin is a cogent option for milia and aforesaid variants. Carbon dioxide and erbium yttrium aluminium garnet lasers demonstrate variable although beneficial results [9,10].

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*Table 1: Classification of cutaneous cysts [4].*
Figure 1: Milia-cyst lined with stratified squamous epithelium, laminated keratin and superimposed epidermis [11].

Figure 2: Milia-cyst coated with attenuated squamous epithelium, keratin impaction and hyperkeratosis of superficial epidermis [12].

Figure 3: Milia-numerous stratified squamous epithelium lined cysts with epidermal invaginations and keratinous lamellae [13].
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Figure 4: Milia-giant cyst with keratin ingress, layering of stratified squamous epithelium and curvy, superimposed epithelium [14].

Figure 5: Milia-cyst with centric aperture, keratin flakes and attenuated, enveloping squamous epithelium [14].

Figure 6: Milia-Sub-epidermal cyst with squamous coating and circumscribing stratified epithelium with hyperkeratosis and acanthosis [15].

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Figure 7: Milia-cyst located in the superficial dermis, stratified epithelial lining and loose keratin [16].

Figure 8: Milia- sub-epidermal cyst with keratinous lamellae, attenuated squamous epithelium, intact granular layer and superficial stratified squamous epithelium [17].

Figure 9: Milia- cyst lined with stretched squamous epithelium, sub-epidermal foci of inflammation and stratified superimposed epidermis [18].

Figure 10: Milia-cyst with keratinous flakes, squamous epithelial lining and dermal inflammatory aggregates [19].

Figure 11: Milia-numerous cysts with stratified squamous and granular cell layer, concentric keratin and encompassing stratified squamous epithelium [20].

Figure 12: Milia-concentric keratinous aggregates, prominent granular cell layer and stratified squamous epithelial lining [20].

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Conclusion

Miniature, superficial, keratin impacted, epidermoid cysts derived from pilosebaceous apparatus or eccrine sweat ducts are cogitated as milia. Morphologically, multiple cysts are layered with stratified squamous epithelium, devoid of adnexal structures and impacted with keratinous lamellae. Multiple epidermal cysts and milia are superimposed upon sites of previous herpes zoster infection, thereby demonstrating Wolf’s isotopic response along with several genodermatosis which can concur with milia. Milia necessitate a demarcation from eruptive syringoma, miliaria crystallina, eruptive vellus hair cyst and generalized milia like idiopathic calcinosis cutis. Symptomatic milia can be appropriately cauterized. Administration of oral etretinate is beneficial in multiple eruptive milia. Topical application of the tretinoin cream, oral azithromycin, carbon dioxide and erbium yttrium aluminium garnet lasers demonstrate efficacious results.

Bibliography


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11. Image 1 Courtesy: Basic medical key.


15. Image 6 Courtesy: Internet Scientific Publications.

16. Image 7 Courtesy: Online resources for healthcare professionals.

17. Image 8 Courtesy: Core.ac.uk.


20. Image 11 and 12 Courtesy: eScholarship.

21. Image 13 Courtesy: JAAD.

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