Case Report

Congenital Epidermal Inclusion Cyst On the Breast: A Case Series of a Rare Entity

Abstract
Epidermal inclusion cysts (EICs) are the most common cutaneous cysts, occurring anywhere in the body but infrequently on the breast. It is usually seen in the third and fourth decades of life and very rarely seen in children. They can be either congenital or posttraumatic. Most of the EICs described in children were secondary to trauma. Congenital EIC on the breast is quite a rare entity. Thus, we herein describe two cases of congenital EIC on the breast in two infants of 3 months and 9 months of age, which to the best of our knowledge has not been reported previously.

Keywords: Breast, congenital, epidermal inclusion cyst

Introduction
Epidermal inclusion cyst (EIC) represents the most common keratinous cyst of the skin. Several reports of posttraumatic EICs are available, and mostly, they occur on trauma-prone sites of the body, i.e., hands and feet.[1-3] In fact, only a few cases of breast localization has been reported in the literature so far, the vast majority being posttraumatic.[1] Here, we have reported two cases of congenital EIC on the breast in 3-month-old and 9-month-old infants.

Case Reports

Case 1
A 9-month-old male child presented to the outpatient department, with the complaint of single, asymptomatic, slowly growing white-colored lesion on his right breast since birth. His left breast was normal. There was no history of trauma, pain, itching, and discharge from the lesion or nipple. General physical examination and systemic examination were unremarkable.

On cutaneous examination, there was a single pearly white dome-shaped lesion of 0.8 cm × 1 cm present on the right areola in vicinity of right nipple. On palpation, the lesion was soft, fluctuant, and nontender [Figure 1a]. The mass was excised in toto and expressed thick foul-smelling cheesy material. On histopathological examination, a cyst was seen within the dermis which was lined by stratified squamous epithelium with granular layer and its lumen consisted of keratinous material. No adnexal structures were observed [Figure 1b]. Thus, a diagnosis of EIC was made.

Case 2
A 3-month-old male infant presented with an asymptomatic, gradually progressive single white-colored lesion on the nipple of right breast since birth [Figure 2a]. Left breast was normal. There was no history of infection, trauma, pain, itching, and discharge from the nipple. General physical and systemic examination was within normal limits. On cutaneous examination, there was a dome-shaped mass of 1 cm × 2 cm present on the right nipple. On palpation, the lesion was freely movable, nontender and had a smooth surface. There was no evidence of keratin-filled punctum over the lesion. Under general anesthesia, total excision of cyst was done. Histologic examination revealed that the cyst wall was lined with stratified squamous epithelium with a granular layer, which was filled with keratinous material and seen within the dermis [Figure 2b]. Thus, the final diagnosis of EIC of the breast was made. There was no evidence of recurrence during the 1 year follow-up period after excision in both the cases.

Discussion
EICs are the type of epidermal cysts which arise due to sequestration of epidermal cells seen within the dermis which was lined by stratified squamous epithelium with granular layer and its lumen consisted of keratinous material. No adnexal structures were observed [Figure 1b]. Thus, a diagnosis of EIC was made.

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in the dermis, which may occur either congenitally or due to posttraumatic implantation of epithelial cells in the dermis.\(^1\)

The exact pathogenesis of sequestration of epithelial cells is not known. Posttraumatic implantation and sequestration of epithelial cells following penetrating and blunt trauma can be explained; however, congenital epithelial cell sequestration and factors associated with it have not been clearly explained. Epstein and Kligman\(^2\) in their study on epithelial cysts have mentioned that pluripotent stem cells, rather than mature buried epithelial cells, play an important role in the cyst formation and differentiation.

Several reports of post-traumatic EICs are available and mostly they occur on trauma prone sites of the body, mainly on the fingers, toes, hands and feet. They are also reported on other sides secondary to trauma following surgical incisions and penetrating trauma due to other causes. Breast is the uncommon site for EICs and in most cases, it is seen in adults following penetrating or blunt trauma either post-procedural or secondary to accidental injuries. Considering the paediatric age group, there is only a single case report of post traumatic EIC in a 15 month old girl.\(^3\) To best of our knowledge, there is no case report of congenital EIC on breast. This might be the first case series of congenital EIC on breast.

In our case, lesion was present since birth and was milky white-colored, firm, pin-head-sized lesion. Being superficial, they appear yellow or white in this area. The most common differential diagnosis is milium. Primary milia of the nipple-areola complex are a rare entity, and so far, only one case of the primary milium of the nipple and four cases of the primary milium of the areola have been reported.\(^4\) Clinically, milia are smaller in size, usually not more than 1–2 mm in diameter and reveal round keratinous content on evacuation with needle. EICs, on the other hand, can be of any size but are usually larger than 5 mm in size and grow rapidly to increase in size and express foul-smelling thick cheesy material. However, both EICs and milia share similar histopathological features as both show a cystic space filled with keratinized granular contents and are lined by several layers of thick stratified squamous epithelium.\(^1\) On the basis of origin and histopathological findings, milia and EICs can be considered as similar entities with different clinical spectrum. Thus, congenital small lesion with gradual progression, large size of about 1 cm, cystic consistency with the presence of compressibility, well-defined cyst walls on excision, and expression of foul-smelling cheesy content prompted us to make the clinical diagnosis of congenital EIC of areola.

EICs usually follow benign course, but complications such as inflammation, secondary infection, and abscess formation are not uncommon. Sometimes, cysts grow rapidly, undergo spontaneous rupture, and can undergo ulceration and sinus formation. Rarely, malignant transformation of squamous cell carcinoma in EICs of longer duration has been reported.\(^5\) Surgical excision is the treatment of choice with complete removal of cyst wall to prevent recurrence. Recently, treatment of EICs with erbium-doped yttrium aluminum garnet (Er:YAG) laser fenestration has been successfully tried as an alternative to surgical treatment.\(^6\)

Thus, the objective of reporting this case is to highlight the rare possibility of congenital EIC on nipple-areola complex. In its early stages, it can be confused with the primary milia of the nipple-areola complex as in our case, but it can be differentiated on the basis of clinical findings and histopathology. It is very important to distinguish the two conditions as milia need only simple needling and extrusion, while EICs require complete surgical excision to prevent recurrences and future complication.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and
other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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