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Case Reports of Cutaneous Ciliated Cysts in Paediatric Population, Unveiling the Unusual

Dr Nisa Maria T[1], Dr Ruby Elizabeth[2], Dr Laila Raji N[3]

[1]Junior Resident, Department of Pathology, Government Medical College, Thiruvananthapuram, Kerala, India
nisamariat[at]gmail.com
[2]Assistant Professor, Department of Pathology, Government Medical College, Thiruvananthapuram, Kerala, India
[3]Professor and Head of the Department, Department of Pathology, Government Medical College, Thiruvananthapuram, Kerala, India

Abstract: ***Cutaneous ciliated cysts are rare benign lesions occurring in the lower extremities of young females possibly of Mullerian origin. We present two pediatric cases of cutaneous ciliary cysts in unusual sites i.e. a seven months old girl presented with swelling over back for three months and a five years old girl with swelling over suprasternal region. On examination both lesions were non-tender and present in the subcutaneous plane. Histopathological examination assisted by immunohistochemistry confirmed a diagnosis of cutaneous ciliated lesion possibly of eccrine origin that showed positivity for epithelial membrane antigen.***

Keywords: Cutaneous ciliary cysts, Eccrine origin, Epithelial membrane antigen

1.Introduction

Cutaneous ciliated cyst (CCC) is a benign cystic lesion that often presents as a palpable subcutaneous mass without any symptoms (1). It was originally described as a painless cyst mostly occurring over the lower limbs of young females, between the ages of 15 and 30 years (2).

The location of the mass can be associated with its origin (3). Likewise, the morphological similarity between the lining cells and the Fallopian tubes epithelium suggests their Müllerian origin which is confirmed using immunohistochemical stains including oestrogen receptor (ER) and progesterone receptor (PR), suggestive of Mullerian heterotopia (1). There were few newer hypotheses suggesting eccrine origin and cloacal membrane origin. The eccrine metaplasia hypothesis was proposed following the identification of morphologically similar cysts in male patients with ciliated metaplasia of eccrine glands while the perineal location of some tumours suggested their embryonic origin from cloacal membranes (4).

Outliers of cutaneous ciliated cysts in unexpected locations such as the abdominal wall, back, finger and scapular region have been reported. We report two cases of cutaneous ciliary cyst among paediatric population at unusual sites including back and suprasternal region.

2.Case Report

A seven months old girl child presented with swelling over the back for three months. She had complaints of purulent discharge from the swelling and was on oral antibiotics. On examination, a 2 x 2 cm swelling was noted on the right side of back in the subcutaneous plane with a black punctum. The lesion was excised in toto. We received a skin attached nodular tissue measuring 1.2 x 1.5 x 1 cm which on cut section showed a cyst filled with whitish material.

The second case was that of a five years old female child with swelling over suprasternal region since birth which was gradually increasing in size. We received a skin attached nodular tissue measuring 1 x 0.8 x 0.5 cm and cut section was grey – white.

Microscopy of both showed skin with normal epidermis and underlying dermis showing cystic spaces intervened by fibro-collagenous stroma showing dense mononuclear inflammatory infiltrate. The cyst wall was lined by ciliated columnar epithelium with pseudostratified areas and high-power magnification revealed fine cilia lining the luminal side of the epithelium. Immunohistochemical staining showed positive epithelial membrane antigen (EMA) in the epithelial component while immunoreactivity for ER and PR were negative. Final diagnosis was consistent with cutaneous ciliated cyst.

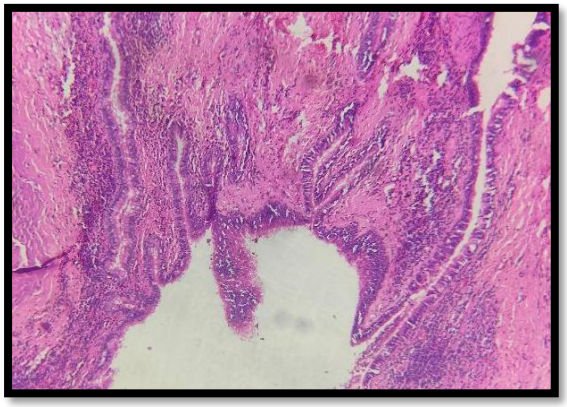

Figure 1: Low power magnification showing cystic spaces intervened by fibrous septae showing mononuclear inflammatory infiltrate (H&E. x 100)

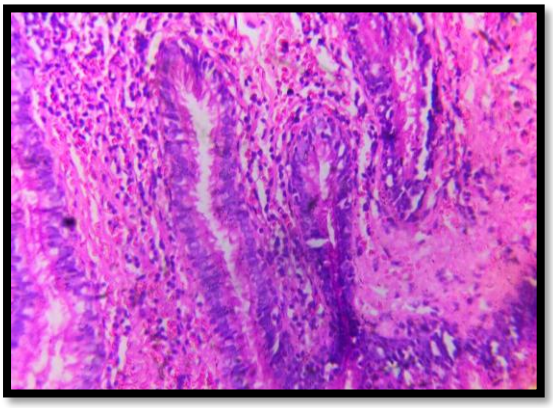

Figure 2

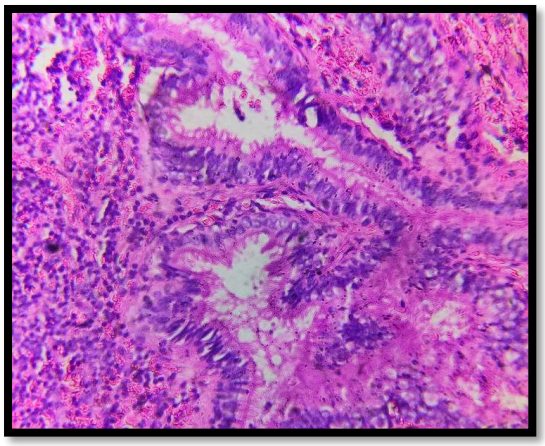

Figure 3

Figure 2 and 3: High power magnification of cyst lined by ciliated columnar epithelium with focal areas of pseudostratification (H&E x 400)

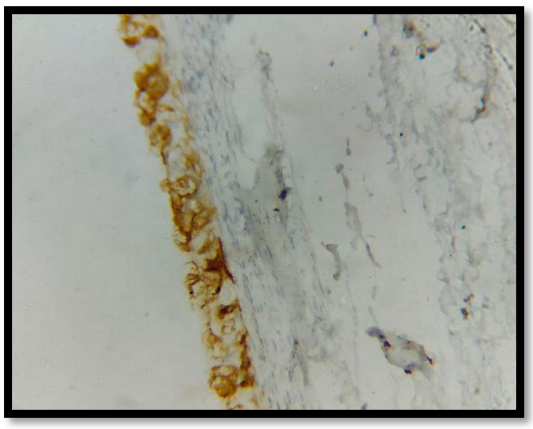

Figure 4: Immunohistochemical test for Epithelial Membrane Antigen showing diffuse positivity

3.Discussion

Cutaneous ciliated cysts are exceptionally rare, benign cystic lesions that occur predominantly in young females, with less than 60 cases reported in the literature till 2015 as cited by Anbarserry et al. (1) and these lesions were found in the lower extremities of child-bearing age with 50 cases in females and only 10 cases in males. However, there have been a small amount cases reported in other anatomic locations and in male cohorts (2). It has been previously suggested that cutaneous ciliated cysts with oestrogen and progesterone receptor positivity are best termed cutaneous Mullerian cysts, while lesions negative for these receptors should be referred to as cutaneous eccrine cysts.

But in the present report, the lesion was seen in rare locations including back and suprasternal region. Also, the absence of ER and PR with positivity for EMA suggests an eccrine origin for this neoplasm which is usually encountered in male patients. Typically, these lesions are slow-growing and may be painful or painless depending on location and size. Another point favouring eccrine origin from foetal eccrine ducts is the presence of ciliated epithelial cells (4). In 1982, Leonforte (5) studied the similarities and relationship of the cyst to the sweat glands and described two features of apocrine sweat glands seen in these ciliated cysts: either the presence of PAS-positive granules or epithelial apical caps. Leonforte (5) considers the epithelial change which appears similar to fallopian tube as metaplastic change due to chronic irritation of the pluripotent cells.

Differential Diagnosis

Few differentials which can be considered for CCC include branchial cleft cyst, teratoma, bronchogenic cyst and thyroglossal cyst. However, the location of the lesion in the lower extremity and the absence of mucous gland, together with the immunohistochemical positivity for EMA and negative ER, PR, CEA confirms the diagnosis of cutaneous ciliated cyst possibly of eccrine origin (1).

Management

The recommended treatment for this cystic lesion is surgical removal for which recurrence after surgery has not been reported in the literature as in our cases while conservative treatment is usually applied in asymptomatic cases (1). Surgical excision is the “gold standard” therapeutic approach in symptomatic, complicated cases or atypical localizations, like cyst close to large vessels. Other treatment modalities include percutaneous aspiration or surgical cyst excision by thoracotomy, sternotomy, or video-assisted thoracic surgery. The prognosis is excellent after resection with only one case of recurrence reported in literature.

4.Conclusion

Cutaneous Ciliated Cysts are rare and generally they are clinically asymptomatic presenting commonly among young females. The location of this lesion is mostly associated with its origin with majority of them presenting as cutaneous Mullerian cysts in the lower limbs and rare cases showing eccrine origin. Outliers of CCC are expected in unusual locations like back, abdominal wall and finger. In the present study, the CCC was diagnosed in paediatric cases. Treatment varies from conservative management to surgical excision, depending on the symptoms. With proper management, patients with

cutaneous ciliated cysts usually have an excellent prognosis and quality of life.

References

[1] Anbarserry I., Board S., Alhamar M., Almousa A., Board S., Alsamarrae O., Qader M.A. and Alsindi K. Cutaneous Ciliated Cyst. Bahrain Med. Bull. 2018: 40(4): 240 – 241.
[2] Hill Z., Stone R., Weiner R. and Buckley K. A rare case of a cutaneous ciliated cyst of Mullerian origin found in the lower extremity: A case report. Foot and Ankle Surgery: Techniques, Reports & Cases. 2023: 100270.
[3] Kim Y.H. and Lee J. Cutaneous Ciliated Cyst on the anterior Neck in Young Woman: A Case Report. World J. Clin. Cases. 2020: 8(19): 4481-4487.
[4] Kim Y. and Kim H. Case Report: The Cutaneous Ciliated Cyst in Young Male: The Possibility of Ciliated Cutaneous Eccrine Cyst. Hindawi Publishing Corporation Case Reports in Medicine. 2015: Article ID 589831.
[5] Leonforte J F. Cutaneous Ciliated Cystadenoma in a Man. Arch. Dermatol. 1982: 118(12): 1010-1012