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Case Report

Congenital umbilical epidermal inclusion cyst masquerading as raspberry tumor– A common cyst in rare site

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ABSTRACT

Introduction: Umbilical tumours or lesions are rare; therefore, it is of outmost importance to navigate every umbilical nodule or growth cautiously. Only 10 cases have been reported to date. The present case has been diagnosed as congenital EIC in the umbilicus, the first time this has been reported in the archives.
Case Report: A 5 year old female child with umbilical nodular swelling since birth presented to the surgery OPD.A clinical diagnosis of "umbilical adenoma" was made. An excisional biopsy with abdominoplasty was done under general anaesthesia, and the specimen was submitted for HPE. On gross examination, we received an umbilical stump measuring 1 x 1 x 0.4 cm. A raised, firm nodular swelling was noted on the umbilical knot along with keratin debris. On cut, it showed a well-circumscribed, tiny gray-white nodule measuring 0.5 x 0.4 cm. On microscopic examination, a diagnosis of "Umbilical Epidermal Inclusion Cyst" was given.
Conclusion: Umbilical EIC is a rare entity that may present as an umbilical nodule and is detected in every age group with a female preponderance. Underlying etiopathogenesis can be congenital, idiopathic, or post-iatrogenic. Over 80% of the umbilical EIC is located above the fascia. The histopathological examination is the only modality that leads to an exact diagnosis and is considered the "gold standard" for detection of commonest cyst occurring in the rare site.

1. Introduction

Umbilical tumours or lesions are rare; therefore, it is of outmost importance to navigate every umbilical nodule or growth cautiously to rule out the possibility of an embryological remnant and associated congenital anomalies in infants and children, or hidden malignancy in adults.[1]

Literature attests to the rarity of umbilical epidermal cysts (EIC); to the best of our knowledge, only 10 cases have been reported to date. The present case has been diagnosed as congenital EIC in the umbilicus, the first time this has been reported in the archives.

2. Case Report

A 5 year old female child with umbilical nodular swelling since birth presented to the surgery OPD. The swelling was not associated with pain, fever, or any other symptoms. The child was born through vaginal delivery, and it was uneventful. The prenatal and postnatal courses were fair and clinically insignificant. A clinical diagnosis of "umbilical adenoma" was made. An excisional biopsy with abdominoplasty was done under general anaesthesia, and the specimen was submitted to the histopathology lab for HPE.

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2.1. Histopathology findings

On gross examination, we received an umbilical stump measuring 1 x 1 x 0.4 cm. The outer surface was smooth and rounded. Circumferential skin measures 1 cm in diameter. A raised, firm nodular swelling was noted on the umbilical knot along with keratin debris. [Figure 1] On cut, it showed a well-circumscribed, tiny gray-white nodule measuring 0.5 x 0.4 cm. [Figure 2] Tissue was completely processed; no remnants were kept.

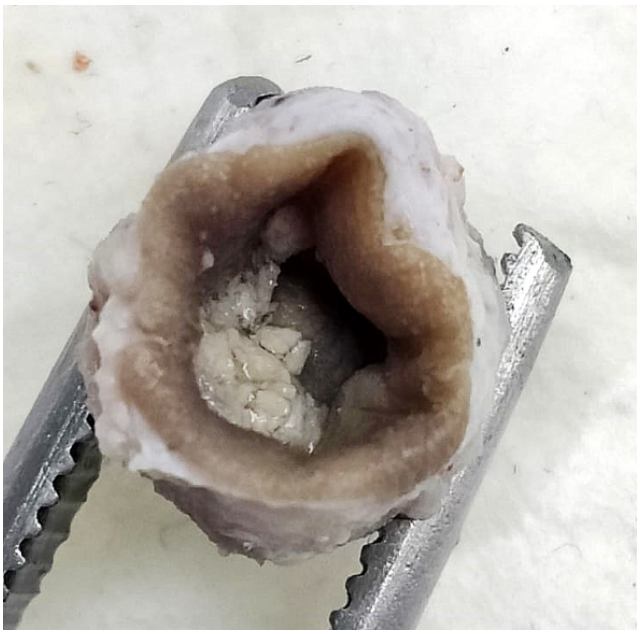

Fig. 1: Gross specimen of umbilical stump show a raised nodule on skin surface of umbilical button.

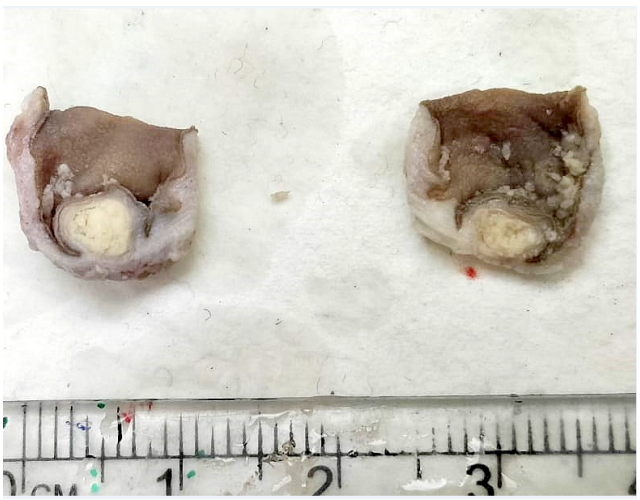

Fig. 2: Cut surface show a well circumscribed tiny grey white nodular growth.

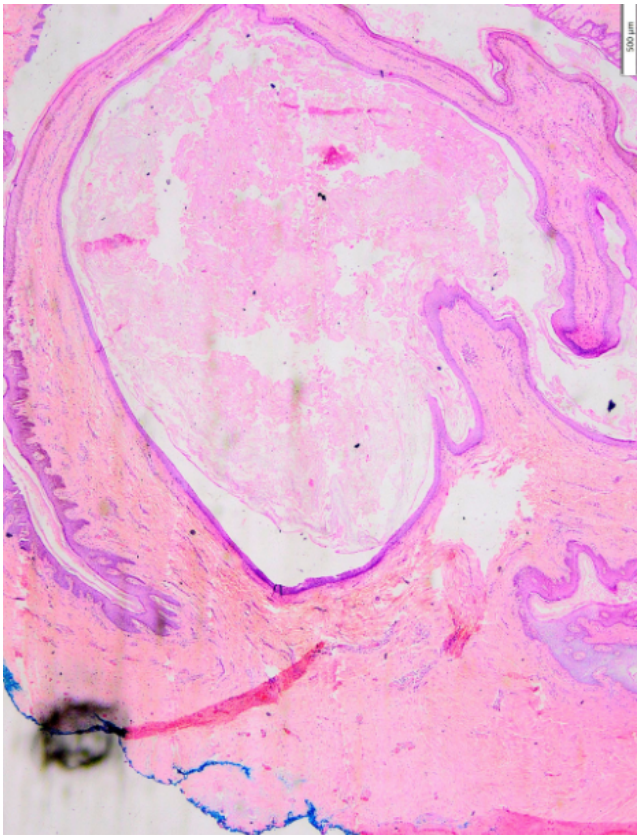

Fig. 3: Cyst lined by stratified squamous epithelial lining with preserved granular layer (H&E, 400X).

On microscopic examination, H&E stained sections are covered with skin, showed hyperkeratosis at places. Underlying dermis showed a well defined cyst lined by stratified squamous epithelial lining with preserved granular layer. [Figure 3] Cyst cavity was filled with abundant keratinous flakes. Adjacent stroma showed adnexal structures and focal mild perivascular lymphocytic inflammatory infiltrate. No evidence of dysplasia or neoplastic pathology seen in the biopsy examined. A histomorphological diagnosis of “Umbilical Epidermal Inclusion Cyst ” was given.

3. Discussion

Before reviewing the literature, we collected all differential diagnoses reported to date at the umbilicus. One should keep all the differential diagnoses in mind while dealing with an umbilical nodule.

There are many congenital umbilical anomalies described in literature, such as infantile umbilical hernia, exomphalos (omphalocele), and many vitelline duct remnants, i.e., vitelline fistula, Meckel’s diverticulum, vitelline sinus, vitelline cyst, vitelline band, mucosal remnants, etc. These congenital anomalies were found to be

associated with certain conditions like Robinow syndrome, renal agenesis-dysplasia, foetal growth retardation, single umbilical artery, hydrops fetalis, anencephaly, monozygous twinning, and achondroplasia[2,3], Down syndrome, prematurity, and mucopolysaccharidosis.[1,3]

Urachal remnants or anomalies include the persistent omphalomesenteric duct, as well as congenital lesions such as the Urachal sinus, Urachal cyst, Urachal diverticulum, Urachal fistula, and patent urachus.

Umbilical granuloma, umbilical polyp, umbilical papilloma, and umbilical tumours are the non-congenital lesions described.[4,5]

Umbilical tumours are commonly found in adults and the elderly. They may be primary or secondary (metastatic).

Benign tumours account for 57% of all umbilical tumors, followed by metastatic deposits.[4,5] Primary benign umbilical tumours are melanocytic tumors, fibroepithelial papillomas, seborrhoeic keratoses, dermatofibromas, neurofibromas, juvenile hemangiomas, keloid, granular cell myoblastoma, desmoids, and lipomas.[4,5]

Primary malignant umbilical tumours are very rare, comprising only 17% of cases, and the spectrum includes melanoma, basal cell carcinoma, and adenocarcinoma. Metastatic umbilical carcinomas are commonly reported, accounting for 83% of malignant cases most commonly primary in gastrointestinal tract.[4,5] Such tumours are called "Sister Mary Joseph's Nodule",[6] and although grossly they are typically less than 5 cm in diameter, they may enlarge to form a protruding tumour. Histomorphologically, it is a metastatic deposit of adenocarcinoma, most often from gastric adenocarcinoma in men and ovarian cancer in women. However, metastases from sarcomas, melanomas, and mesotheliomas have also been reported.[4,6]

Epidermal inclusion cysts refer to the implantation of epidermal cells into the dermis following any kind of trauma, such as surgery. An EIC occurring in the umbilicus is a rare phenomenon.[7]

Mc Clenathan et al.[8] described umbilical EIC in 7 cases for the first time in 2002. Later, single cases were reported in each study performed by Andreadis et al.[9] in 2007, Camenisch et al.[10] in 2012, and Christine Li et al.[7]recently in 2022.

According to the literature,[7–10] the average mean age of presentation is 42 years, observed in wider age group starting from 9 to 61 years Female preponderance is clearly noted with a male-to-female ratio of 1:4. The most common clinical presentations are pain and mass in 70% of cases, erythema in 50% of cases, and discharge in 40% of cases. Symptoms can last anywhere from a day to years. On gross examination, EIC found to be located above the fascia in 80% of reported cases, and below the fascia in the remaining 20%.

The size dimension and cut surface are poorly documented in the literature and radioimaging. Excision of the lesion was considered treatment in 90% of cases. However, 1 patient was given a course of antibiotics.[7–10]

4. Conclusion

Umbilical EIC is a rare entity that may present as an umbilical nodule and is detected in every age group with a female preponderance. Underlying etiopathogenesis can be congenital, idiopathic, or post-iatrogenic. Umbilical EICs presenting as nodules have many other differential diagnoses; however, being the commonest cyst encountered almost daily, EIC may be seen at this rare site. Over 80% of the umbilical EIC is located above the fascia. The histopathological examination is the only modality that leads to an exact diagnosis and is considered the "gold standard" for detection of commonest cyst occurring in the rare site.

5. Conflict of Interest

None.

6. Source of Funding

None.

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