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CASE REPORT

A RARE CASE REPORT OF CUTANEOUS HYBRID CYST FROM INDIA, IS IT TELLING US SOMETHING?

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ABSTRACT

Many types of the cutaneous hybrid cysts are described in literature since they were first described in nineteen sixties. These hybrid cysts are developing from Folliculo-sebaceous apocrine units in the skin. These can be single or multiple. When such type of cyst is detected, these patients are frequently examined for any syndromic association like Lowe syndrome, Gardner syndrome and Gorlin Goltz syndrome. However, literature shows that many of the cases are non-syndromic. There can differentiation towards infundibulum, isthmic region or lower parts like follicular matrix, inner root sheath and hair. Exact pathogenesis involved in the formation of these cysts is largely unknown. Follicular germinative cells are hypothesized to form these types of cysts which are capable of divergent differentiation. There are occasional case reports from India. Hereby we report rare case of cutaneous hybrid cyst in middle aged male with right forearm swelling for 8 months. On histopathological examination lesion, it showed epidermal inclusion cyst and Pilomatricoma. The patient was asymptomatic apart from the swelling. He had no gastro-intestinal problems and his family history was negative for the colorectal carcinoma.

INTRODUCTION

Cutaneous hybrid cysts are rare type of dermal lesions. More common cysts include epidermal inclusion cysts, trichilemmal or pilar cysts. Less common forms are steatocystoma, cutaneous Hydrocystoma and vellus hair cyst. Hybrid cysts are combination of these individual lesions. Epidermal inclusion cyst and Trichilemmal or Pilar cyst is the most common combination found in Hybrid cysts. Other combinations are rare and they have more chances of association with Gardner syndrome or Lowe syndrome. Gorlin Goltz syndrome is also associated with cutaneous cysts like steatocystoma. To the best of our knowledge only occasional case reports of Hybrid cysts are reported from India. Here by we present the case of middle-aged male with asymptomatic swelling on right forearm for the past 6 months. Histopathology showed Large epidermal inclusion cyst with small pilomatricoma. The patient denied history of gastro-intestinal problems and family history of colorectal carcinoma.

CASE REPORT

A middle-aged male came with swelling on the right forearm since last 8 months. The swelling was initially small and slowly grown to size of 1.5 cm in diameter. The swelling was painless and overlying skin did not show redness. Skin was slightly pinchable over the swelling. Punctum and sinus tract was not seen. Swelling was devoid of punctum. Slip sign was negative. There were no similar swellings on the body or history of previous surgery. Patient was well built and nourished. His vitals were stable with pulse of 83 beats per minute. Respiratory rate was 10/min. Hence with adequate sterile precautions, the swelling was excised under local anaesthesia and sent for histopathological examination.

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Grossly one already cut opened cystic tissue measuring 1.5 cm in diameter was received. The wall of cyst measured 0.1 to 0.2 cm in thickness. Lumen of cyst was filled with pultaceous material. There was small firm part attached to cyst wall on its outer aspect measuring 0.3 cm in greatest dimension. The hematoxylin and eosin stained sections showed a cyst wall in dermis. The cyst was lined by stratified squamous keratinising epithelium with intact granular cell layer. Numerous, loose dry keratin flakes are noted to fill the lumen of the cyst. Hair shafts or adnexal structures were not seen. At one area of the circumference of the cyst wall there was attached cutaneous adnexal tiny neoplasm. The neoplasm composed of strips of squamous epithelium with empty nuclear spaces. There were occasional basaloid cells seen. Calcification and fibrosis was noted in the adjacent area. Finally, resection specimen was reported as cutaneous hybris cyst composed of Epidermal inclusion cyst and Calcifying epithelioma of Malharbe. Dermoid cyst was ruled out as there were no adnexalstructures seen. The patient did not give history of altered bowel habits, bleeding per rectum or abdominal pain. There were no other swellings on the body. In the family history, none of the relatives had colorectal carcinoma. The lesion was adequately resected, however patient was counselled regarding the recurrence and syndromic association and advised to have regular follow up for consultation in surgery every three monthly. The surgical site wound healed properly and stitches were removed. There was no infection. Scar was healthy. After 3 months of follow up patient was healthy and there was no recurrence.

DISCUSSION

McGavran et al described the how to differentiate pilar and epidermal inclusion cysts.(1)Also,later on in 1983, Philip et al from Virginia studied cutaneous cysts in Gartner syndrome. They found hybrid changes in the cutaneous epidermal cysts.

As most areas were composed of follicular or infundibular epithelium, focal areas were showing pilomatricoma like changes.

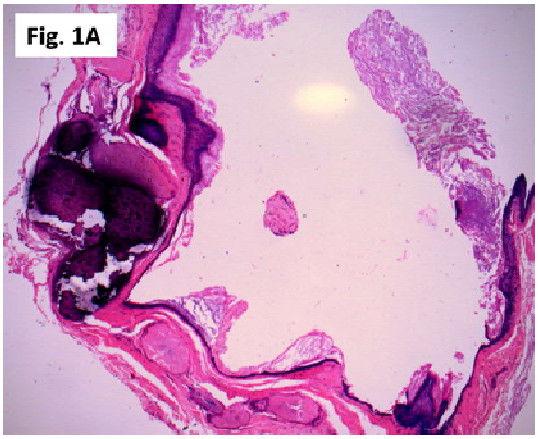

Figure 1A. Hematoxylin and eosin-stained section on scanner view of 20x magnification show cyst lined by keratininsing stratified squamous epithelium. Lumen is filled with Keratin flakes. Adnexal structures or hair shafts are not seen. Wall of cyst also shows many foci of dystrophic calcification and few islands of epithelial cells

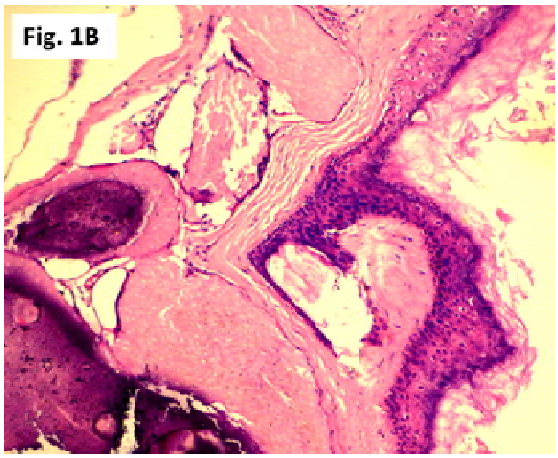

Figure 1B. The Hematoxylin and eosin-stained section on 200x magnification shows Stratified squamous epithelium with intact granular cell layer. There is presence of islands squamous cells with Ghost cell appearance due to empty nuclei. This with foci of calcification is indicative of Pilomatricoma component

These were associated with Gardner syndrome. (2). In the same year Brownstein studies most common hybrid cysts. The upper portion of the cysts was more similar to infundibular or follicular epithelium whereas the lower portion was similar to trichilemmal cysts. This finding reinforced their origin from infundibular region and isthmic region respectively. These cysts may show follicular, sebaceous and apocrine differentiation. (3)Their various combinations were described in the review article by Rquena et al in 1983. Thus, the spectrum was expanded from combination of epidermal and pilar cysts to other lesions arising from the pilosebaceous units. Depending on differentiation from infundibulum, isthmic portion, hair matrix like differentiation, sebaceous gland and duct differentiation various components can be seen. Pilomatricoma arises from lower part of hair unit. Its shows hair matrix like differentiation. Steatocystomashows sebaceous gland and duct differentiation. Vellus hair can be found in cyst to label it as Vellus hair cyst. There can be decapitation of secretions from the inner epithelium in the cyst indication it to be Apocrine hydrocystoma. Morphology of suprabasal acantholysis with corps ronds in acanthotic epithelium of the cysts is consistent with diagnosis of Warty Dyskeratoma. (4)

It is important to diagnose these hybrid cysts as they may be early indicators for the patients with genetic syndromes. Gardner syndrome is autosomal dominant and can manifest with variable penetrance at wide age range. Different manifestations are common in the different age groups. Most common and early features are cutaneous cysts like sebaceous cyst and other subepitheial cysts. Osteomas are common in the bones especially of the skull. Other bones can also be involved. These patients have various manifestations of the soft tissues like lipomas, fibromas and neurofibromas. In gastrointestinal system they have adenomatous polyps which can transform to adenocarcinomas with increasing age. Periampullary carcinoma is another common manifestation. These patients are more prone for the development of desmoid tumours. Other rare manifestations include intracranial tumours, malignant bone tumours like osteosarcoma and chondrosarcoma, papillary thyroid carcinoma. They have increased pigmentation of retina and dental problems like impacted tooth and detigerous cysts. Gardner syndrome shares the same genetic mutation as that of Adenomatous familial polyposis coli. APC gene located on long arm of the chromosome number 5 is commonly affected. Variable penetrance of this gene causes extraintestinal manifestations in the Gardners syndrome. (5)The hybrid cutaneous cysts can help to early diagnose the patient and give him prophylactic treatments.

Lowe syndrome is associated with cutaneous cysts like steatocystoma or occasionally vellus hair cyst. It is oculo-cerebro-renal syndrome due to defect in the phosphatase enzyme causing more release of extracellular lysosomal enzymes and tissue damage. (6,7)Similarly, Gorlin Goltz syndrome which is also known as nevoid basal cell carcinoma syndrome has various cutaneous hybris cysts. Cysts with steatocytoma component are more commonly seen in this syndrome. These cysts have corrugated squamous lining epithelium with absent granular cell layer and corrugated lining. Patchy basal or epithelial staining for K19 and continuous basal staining for BCL2 is common in the syndromic Steatocytoma cysts.(8)Thus, when we get different combination from epidermal and pilar cysts like steatocytoma, pilomatricoma or other less common type of differentiation we need to be more vigilant to search for the underlying syndrome. Many times, the cysts are not associated with syndromes but they present with uncommon combinations and it is necessary to address them. For some cysts just adequate surgical excision is necessary with sufficient and there is no recurrence. Some cyst with the neoplastic nature requires rection with adequate margins to prevent their recurrence. Some may require local radiotherapy also. This was observed in a case report by Diep et al. (9). Our case is rare. This was a solitary Cutaneous Hybrid cyst lesion composed of pilomatricoma and epidermal cyst. Simple excision with adequate surgical margins is curative. Patient denied the family history and any history of gastrointestinal problems. It is advisable for regular follow up to detect and treat other manifestations if they occur.

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