Original Research Paper



General Surgery

CARNEY COMPLEX PRESENTED IN NIPPLES OF A YOUNG FEMALE

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PATIENT PRESENTATION: 19 year female, presented with the chief complaint of bilateral swelling in the breast since 2 years, which was associated with serous discharge. A small swelling in left eyelid since 2 years. On examination, 3x2x2 cm pedunculated mass protruding out from the right nipple areola region, involving nipple. 1x1x1 cm pedunculated mass protruding out from the left nipple areola region, involving nipple. no enlarged axillary lymph nodes found. 0.5 x 0.5 cm mass present over left lower eyelid medial aspect.

DİAGNOSIS: CARNEY COMPLEX: MYXOMA (Biopsy proven)

TREATMENT: Bilateral breast conservation surgery

FOLLOW UP: Patient had no post operative complaints and complications. She was cosmetically satisfied.

KEYWORDS:

INTRODUCTION:-

Carney complex is a rare autosomal dominant disorder. Benign tumors of connective tissue (myxomas) are common in individuals with Carney complex, and, most commonly, are found in the heart. Here is the case of a young female who has myxoma of nipple and eyelid without involving heart and endocrine glands.

CLINICAL HISTORY:-

 $19\,year$ female, presented with the chief complaint of bilateral swelling in the breast since 2 years , which was associated with serous discharge. It was not associated with any pain , bloody discharge. A small swelling in left eyelid since 2 years. On examination , 3x2x2 cm pedunculated mass protruding out from the right nipple areola region, involving nipple. 1x1x1 cm pedunculated mass protruding out from the left nipple areola region, involving nipple no enlarged axillary lymph nodes found. 0.5 x 0.5 cm mass present over left lower eyelid medial aspect.



RIGHT BREAST

LEFT BREAST



LEFT LOWER EYELID

RADIOLOGICAL

- Ultrasonography was suggestive of BIRADS III LESION IN BILATERAL BREAST, which was isoechoic and circumscribed mass.
- (2) X Ray: showed a white colored mass on the right breast.
- (3) 2 Decho (heart): WNL



INTERVENTION: Blateral Breast conservation surgery was performed. Intra operative findings were, on right side 4x3x2 cm pedunculated mass was presented in the areola-nipple region and on left side 1.5x1.5x1 cm mass was present in left nipple areola region. Masses were not involving deep structures of breast.





RIGHT BREAST

LEFT BREAST

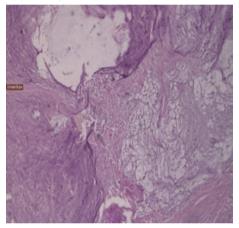
INTRA OPERATIVE PICTURES



GROSS EXAMINATION

CUT-SECTION

HISTOPATHOLOGY REPORT: both the mass shows stratified squamous epithelium lining exhibiting pigmentation at places sub epithelial zone shows fibrocollagenous tissue, mixed inflammatory cell infiltrates proliferating blood capillaries adjoining tissue shows hypocellular dense myxoid stroma with few spindle cells . Features suggestive of MYXOMA.



FOLLOW UP: Patient had no post operative complaints and complications. She was cosmetically satisfied.

CONCLUSION:-

Carney complex without involving heart is very rare condition. In the above mentioned case, patient was having myxoma of nipples and skin involvement of a left eyelid. There was no involvement of adrenal gland, pituitary gland, thyroid, etc and any other skin changes in the body.



FOLLOWED UPAFTER 2 MONTHS

DISCUSSION:

Carney complex is a rare autosomal dominant disorder characterized

by multiple benign tumors (multiple neoplasia) most often affecting the heart, skin and endocrine system and abnormalities in skin coloring (pigment) resulting in a spotty appearance to the skin of affected areas. Carney complex occur due to mutations of the *PRKAR1A* gene, ON CHROMOSOME 17. Benign tumors of connective tissue (myxomas) are common in individuals with Carney complex, and, most often, are found in the heart where they can potentially cause serious, lifethreatening complications including stroke, valvular obstruction or heart failure. A wide variety of endocrine abnormalities potentially can occur in Carney complex affecting a variety of glands.. Less often, myxomas can be found in other areas of the body in addition to or instead of the heart, like breast, nipples , thyroid , adrenal gland, pituitary gland, the eyelids, the external ear canal, etc Almost all people with Carney complex have areas of unusual skin pigmentation. Brown skin spots called lentigines may appear anywhere on the body but tend to occur around the lips, eyes, or genitalia. Myxoma of nipple in young female, can be managed by breast conservation surgery.

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