

Mass on thigh mimicking soft tissue tumor – a rare case report

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Abstract

Hidradenoma papilliferum (HP) is a rare benign, cystic, papillary, apocrine gland tumor that occurs mostly in females on the anogenital area. Rare cases of ectopic (non-genital) HP have been described. These ectopic tumors are seen at various sites and are more common in women. These lesions usually present as asymptomatic slow-growing, red, firm, mobile, well-defined nodule that grows for a long time before resection. The prognosis for HP is very good. Incomplete excision of tumor resulted in tumor recurrence. Malignant transformation is rare. We present a case of 65-year-old man who presented with an enlarging nodule on his left thigh. The histological findings and immunohistochemistry revealed a Hidradenoma papilliferum.

Key words: Benign Neoplasm, Hidradenoma Papilliferum, Apocrine Differentiation, Thigh, Ectopic

Introduction

Hidradenoma papilliferum (HP) is a benign adnexal cystic and papillary neoplasm with apocrine differentiation. Some authors considered this tumor as an analog of intraductal papilloma of the breast [1]. HP is most common in women and only few cases were reported in men [2, 3, 4]. HP is asymptomatic, solitary, slow-growing, skin-coloured nodule, papule or polypoid exophytic lesion, of less than 1 cm in diameter [5]. The tumor primarily affects vulvar, perineal and perianal skin with rare cases being reported in extra-genital or ectopic sites like in head and neck region, axilla, upper limb, back and thigh [2]. The tumor course, clinical and pathological features, treatment and prognosis are similar in both classic and ectopic HP. The diagnosis of ectopic HP can be made only by histopathological examination as they clinically mimic other cutaneous neoplasms.

Case Details

A 65-year-old male presented with asymptomatic papillary growth over left thigh since 2 years. It was initially of a pea size and gradually progressed to a size of 4X4 cms. On local examination it was solitary,

irregular, papillary growth in front of left thigh, 20 cms away from anterior superior iliac spine. It was non tender, skin over swelling was not pinchable and surface over swelling was nodular. No regional lymphadenopathy was present. There was no history of any neurological or skeletal abnormality. Patient is positive for hepatitis B. The routine hematological and ultrasonographic tests were normal.

Gross Findings: The skin excision biopsy of growth revealed skin covered soft tissue mass measuring 8X4X3 cms. The cut section showed a grey white area measuring 3X2X2 cms, with cystic change containing hemorrhagic fluid and fatty areas (figure 1).

Microscopy Findings: Multiple sections studied showed thinned out epidermis and tumor tissue in mid dermis with no connection to overlying epidermis (figure 2a). Tumor tissue is arranged in lobules (figure 2b) and cystic areas (figure 2c). Lobules show cystically dilated ducts with eosinophilic secretions in lumen and papillary projection in to lumen lined by two rows of tumor cells with columnar cells towards the lumen and outer cuboidal cells. Decapitation was seen in the luminal columnar cell layer. Nuclei are monomorphic and vesicular (figure 2d). Few areas showed clear cell change. Adjacent areas showed

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dilated and congested blood vessels. The possibilities of PECOMA, vascular tumor and adnexal tumor with

apocrine differentiation possibly hidradenoma papilliferum was made on histological examination.

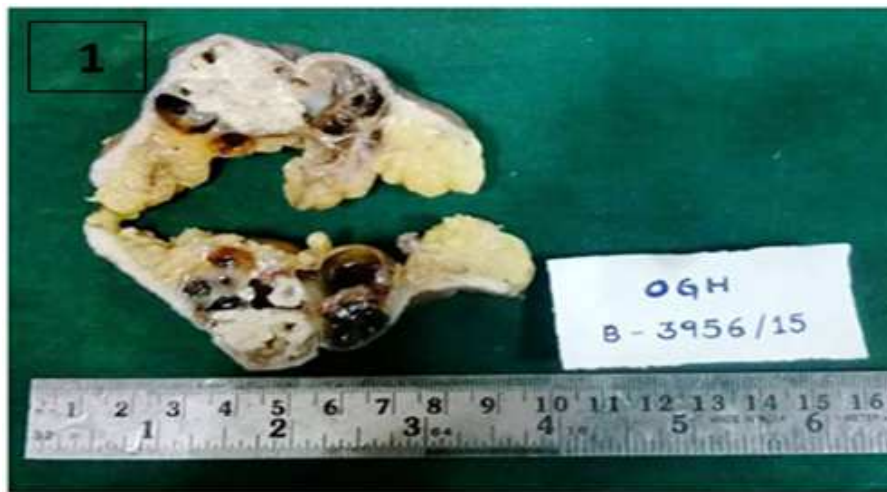


Figure: 1: Gross specimen showing grey white solid area measuring 3X2X2 cms along with hemorrhagic fluid filled cystic areas and fatty areas.

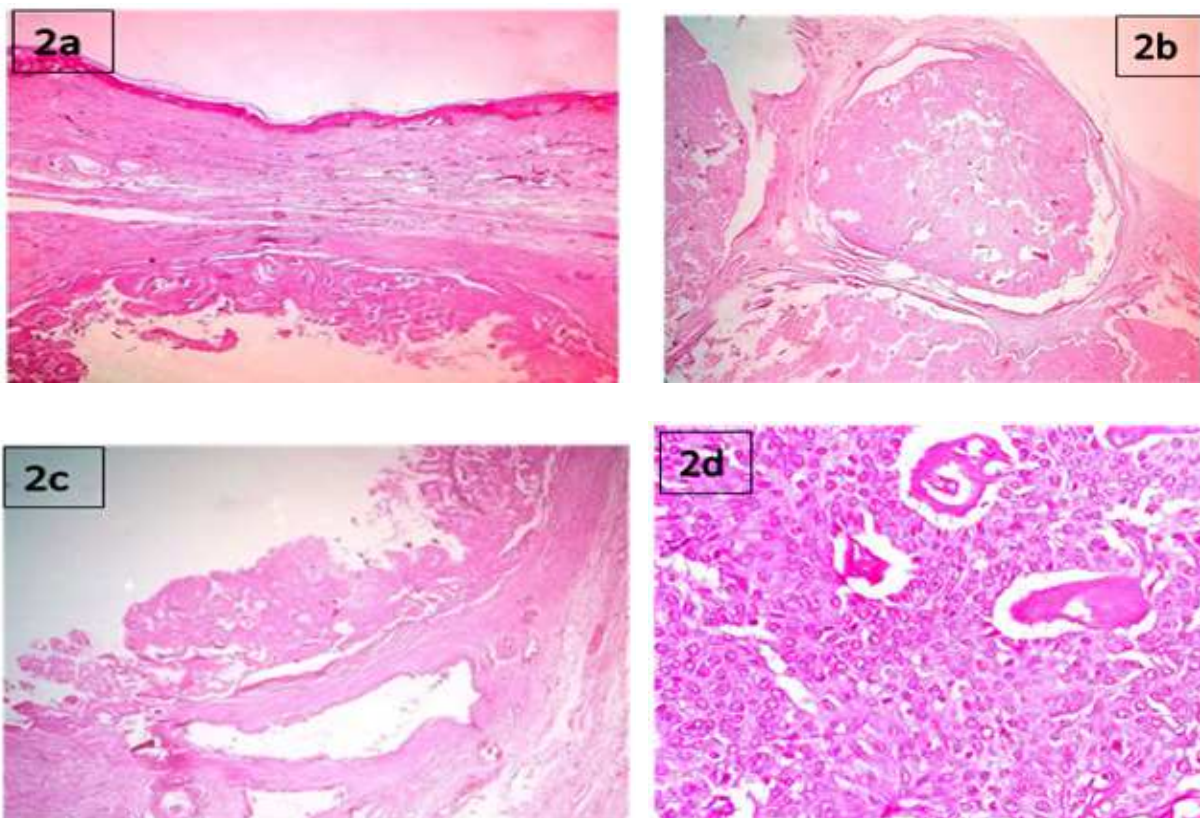


Figure 2a): Hematoxylin-eosin stain (10x) showed the presence of tumor in dermis; **2b)** Hematoxylin-eosin stain (10x) showed tumor tissue arranged in lobular pattern; **2c)** Hematoxylin-eosin stain (10x) showed cystic areas; **2d)** Hematoxylin-eosin stain (40x) showed monotonous cells with central vesicular nuclei.

Further study with Special stain: Periodic acid Schiff (PAS) stain revealed positivity in the eosinophilic secretions (figure 3).

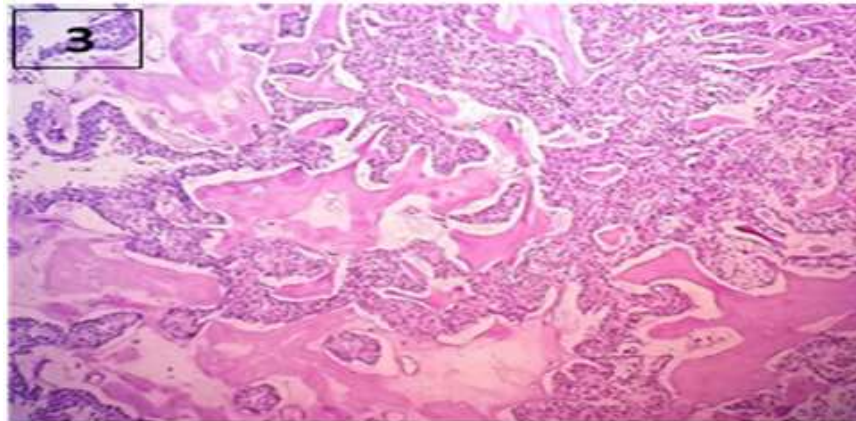


Figure 3: Periodic Schiff stain (10X) showed positivity in secretions.

Immunohistochemistry: Immunohistochemistry (IHC) stain using epithelial membrane antigen (EMA), pan CK, CK5/6, S100, smooth muscle antigen (SMA), CD31, CD34, Her 2 neu, HMB 45, neuron specific enolase (NSE) and Ki67 revealed pan CK, CK5/6, EMA strong positivity in both columnar and basal cells (figure 4a, 4b, 4c). Focal positivity of NSE was seen in tumor cells. SMA, S100, CD 31 (figure 4d), CD 34, HMB 45, her2neu, NSE were negative in tumor cells. Ki 67 was showing focal positivity in <10% of tumor cells. These findings ruled out PECOMA and vascular tumors and confirmed the diagnosis of hidradenoma papilliferum of apocrine origin. There was no evidence of malignant transformation.

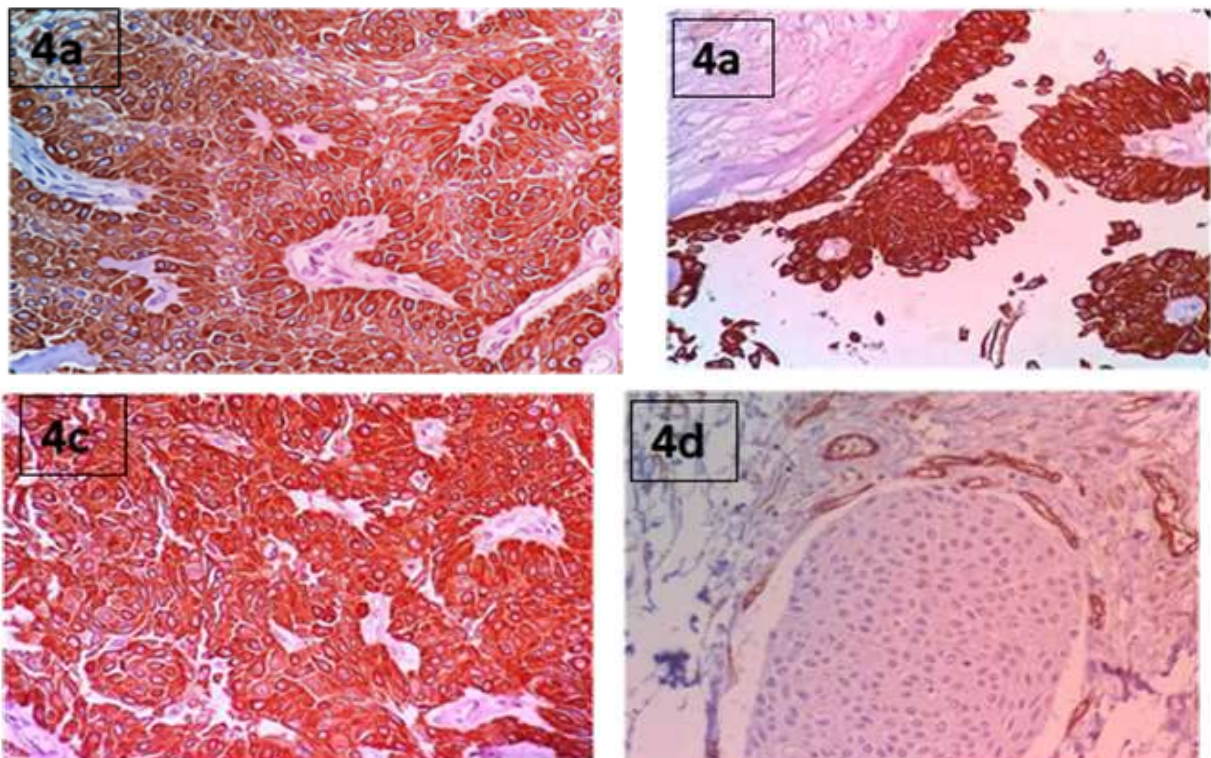


Figure 4a & b): Immunohistochemistry (40X) showed cytokeratine positivity in tumor cells; 4c) Immunohistochemistry (10X) showed ck5/6 positivity in tumor cells; 4d) Immunohistochemistry (40X) showed CD 31 negativity in tumor cells.

Discussion

Hidradenoma papilliferum is an uncommon benign tumor originated in the apocrine glands, which are mainly concentrated in the anogenital and peri-anal regions.

Extragenital localization of tumor called ectopic HP is very rare. 17 cases of ectopic HP were reported so far [2]. The most frequent location has been the head and

neck area followed by the upper and lower extremities. The age range reported is between 8 to 78 years.

HP presents as solitary, firm, well demarcated nodule. These lesions are usually asymptomatic. Pain, pruritus, bleeding or discharge occurs especially if it ulcerates. It is typically smaller than 1 cms, but reports of tumor up to 4 cm have been published [6]. As these tumors clinically mimic other neoplasms such as basal cell carcinoma or metastatic deposits, histological examination is required for the correct diagnosis.

Microscopically, the tumor is characterized by a well circumscribed cystic lesion in the mid dermis with the epidermis being normal, acanthotic, or ulcerated. The tumor may sometimes show continuity with the epithelium [7]. The cystic spaces show papillary folds projected from the cyst wall with eosinophilic material in the lumen. Tumor epithelium is bilayered with a luminal layer of columnar cells showing decapitation secretions and a basal layer of cuboidal cells. The important differential diagnosis of HP is syringocystadenoma papilliferum as both originate from apocrine glands [2].

Syringocystadenoma papilliferum occurs in head and neck region, either denovo or with in nevus sebaceous. It shows duct and cyst like cavities of various sizes extending from the epidermis. These extensions typically have squamous epithelium near the surface of the lesion and is contiguous with a double layered epithelium in the deeper portions of tumor [8]. Histologically, in contrast with the syringocystadenoma papilliferum, HP originate in the mid-dermis, show more complex papillary growth pattern and lacks epidermal surface invaginations. The papillary structures in the HP does not have connective tissue and plasma cell infiltrates [9]. Lesions with features of both have been described. Special stain with Periodic acid Schiff stain showed positive granules in the apices of apocrine cells and secretions in the lumen. Immunohistochemical studies demonstrated that epithelial cells lining the papillations show low-molecular weight cytokeratins positivity. The columnar cells lining the luminal border of tubular structures is also stained by epithelial membrane antigen (EMA), carcinoembryonic antigen (CEA) and gross cystic disease fluid protein-15 (GCDFP-15). S-100 and high-molecular-weight keratins are negative in the tumor cells [10]. SMA positivity is normally the feature of myoepithelial cells but HP demonstrate positivity in focal basal cells thereby indicating the immaturity of

the tumor. Neoplastic epithelial cells lining tubules and papillations also express strong immunoreactivity for androgen and oestrogen receptors.

Local excision is the treatment of choice. Prognosis is good. Incomplete excision attributes for recurrence of the lesions and there is no report of recurrence for the ectopic form [6]. Malignant transformation is not documented in the ectopic HP unlike anogenital hidradenoma papilliferum [6,11].

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