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Case Report

Oncology Section

Syringocystadenoma Papilliferum: An Unusual Presentation

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ABSTRACT

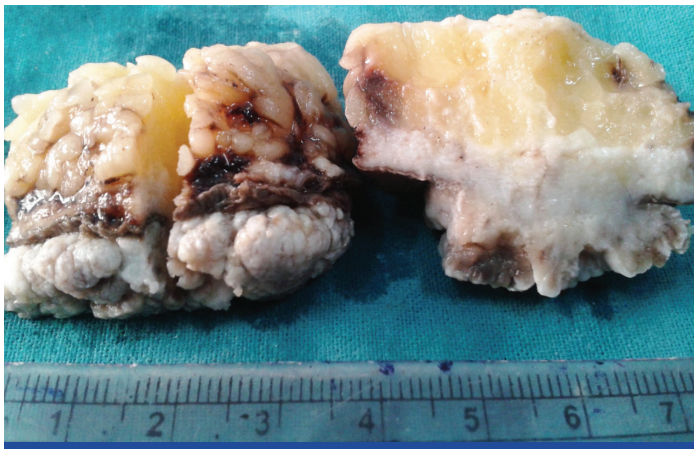
Syringocystadenoma papilliferum is a benign hamartomatous adnexal tumour. Most of the patients present with solitary lesions in the head and neck region at birth or in early childhood. Multiple lesions are rarely seen and those which arise outside the head and neck region are even more uncommon. A case of syringocystadenoma papilliferum with multiple verrucous lesions and which was located in the right flank, an unusual site, has been presented.

Keywords: Syringocystadenoma papilliferum, Abdomen, Multiple, Verrucous lesions

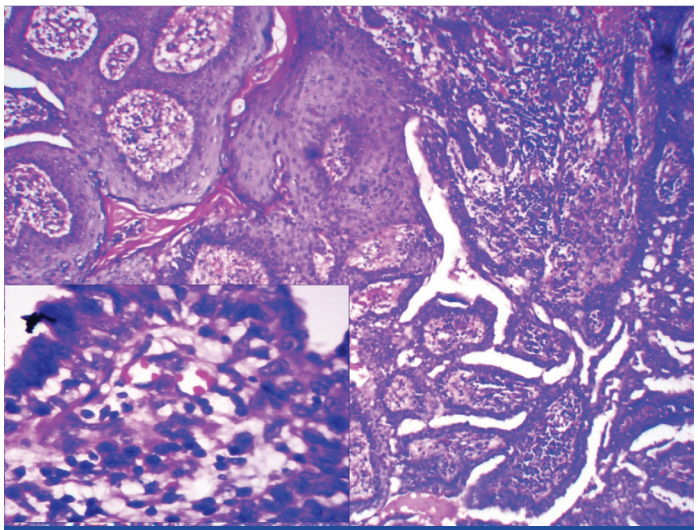
CASE REPORT

A 35-year-old male, a farmer by profession, presented with multiple progressive growths in the right flank, of two year's duration. It had started as plaques for which he had applied traditional topical ointments. The lesions developed a change in texture later and they grew in size. There were no associated systemic symptoms. On examination, multiple papules and nodules were observed on the right lower abdomen, the largest of which measured 5 x 4 cm. The outer surfaces of some of the lesions were eroded and there was a yellowish, creamy, foul smelling discharge. Culture of the discharge from the lesions grew *Pseudomonas aeruginosa* and he was treated with a combination of Ceftazidime and Gentamicin for 2 weeks. Thereafter, the lesions were surgically excised and tissue was sent for a histopathological examination.

Gross examination showed a greyish-white, soft-tissue piece which was covered with yellowish crustations. It measured 4.5 x 3 x 3 cm in dimension. There were papillary projections on the outer surface. Cut-section was homogenous [Table/Fig-1]. Microscopic examination showed acanthotic and papillomatous epidermis with multiple cystic, papillary, and ductal invaginations which extended into the dermis. These invaginations were lined by double layers of cells which consisted of an outer layer of cuboidal cells and a luminal layer of tall columnar cells [Table/Fig-2]. A decapitation secretion was seen in the luminal layer, which stained positive with Periodic acid Schiff's stain. The connective tissue core was filled with plasma cells [Table/Fig-2], few lymphocytes and dilated capillaries. The histological findings were consistent with the diagnosis of syringocystadenoma papilliferum. The post-operative period was uneventful, the patient on regular follow-up and is doing well.



[Table/Fig-1]: Gross picture of the excised tissue. Note multiple papillary projections from the outer surface



[Table/Fig-2]: Photomicrograph showing papillary projections lined by double epithelial layers and lumen filled with dense plasma cell infiltrate (Inset) (H& E x 400)

DISCUSSION

Syringocystadenoma papilliferum presents as brownish or erythematous papules, nodules, or smooth, hairless plaques of varying sizes which range from 5 to 160 mm. Their surfaces can be smooth, flat, papillomatous, or verrucous. Increase in their sizes, crustations, nodular or verrucous transformations are noted at puberty. Most of the patients present with solitary lesions. Multiple lesions are usually associated with naevus sebaceous. Both sexes are equally affected [1].

A majority of the reported cases had lesions in the head and neck region [1,2]. Mammino et al., reviewed 145 cases of syringocystadenoma papilliferum and found 108 cases (75.0%) which showed involvement of the head and neck region, 29 cases (20.0%) which showed involvement of the trunk, and 8 cases (5.0%) which showed involvement of the extremities [2]. Uncommon sites of occurrence which have been reported in the literature include chest, arms, breast, eyelids, axilla, scrotum, thigh, lower legs, toes, inguinal and perineal regions [3-6]. Ninety percent of the cases

show involvement of anatomic sites which are normally devoid of apocrine elements [1]. Nodular variety shows more involvement of the trunk [7].

Few lesions which are reported to be associated with syringocystadenoma papilliferum include viral warts, naevus sebaceous, linear naevus verrucosus, naevus comedonicus, apocrine poroma, apocrine hidrocystoma, tubulopapillary hidradenoma, hidradenoma papilliferum, papillary eccrine adenoma, verrucous carcinoma, apocrine acrosyringeal keratosis, poroma folliculare, linear naevus verrucosa, atypical fibroxanthoma, clear cell syringoma, basal cell epithelioma, sebaceous epithelioma, trichoepithelioma and verruca vulgaris [6-10]. Its association with malignant tumours such as verrucous carcinoma, basal cell carcinoma, sebaceous carcinoma and ductal carcinoma is also known. Syringocystadeno carcinoma papilliferum can also arise in benign lesions [11]. The commonest association is its occurrence within a naevus sebaceous [1].

Histopathology frequently shows multiple cystic invaginations in a background of fibrous tissue. The upper regions of the cystic invaginations are commonly lined by keratinizing squamous epithelial cells which are similar to those seen on the surface epidermis, whereas the lower region contains numerous papillary projections of variable shapes and sizes, which extend into the lumina of the invaginations. These papillary projections are the most characteristic pattern. The glandular epithelium is lined by two layers of cells, namely, tall, columnar cells with oval nuclei and faintly eosinophilic cytoplasm seen at the luminal surface and small cuboidal cells with round nuclei and scanty cytoplasm seen at the base. In some areas, the cells of the luminal layer are arranged in multiple layers and they form a lace-like pattern, resulting in multiple, small, tubular lumina. Decapitation secretion or “apical snouts” are often seen at the luminal surface. Another diagnostic feature of this neoplasm is the presence of a mononuclear inflammatory infiltrate which consists mainly of plasma cells of IgG and IgA class in the fibrous tissue of the papillary projections, alongwith dilated capillaries. Diastase resistant Periodic acid-Schiff (PAS) positivity favours the apocrine differentiation of this tumour [6]. Tumour cells also may show positivity for carcinoembryonic antigen (CEA) and protein-15 [1].

Clinically, the differential diagnosis which is thought of includes cutaneous tuberculosis and pyogenic granuloma. Based on the strong Mantoux reaction which was seen and the high incidence of tuberculosis which is seen in our country, warty tuberculosis was considered as a possibility, but absence of granuloma excluded it. Pyogenic granuloma was considered, based on its soft, exuberant appearance with an eroded surface. However, the absence of a pedicle and relatively scanty bleeding did not favour this diagnosis. Syringocystadeno carcinoma papilliferum, a malignant counterpart of syringocystadenoma papilliferum, is characterized by solid areas and presence of neoplastic cells. There were no features which were suggestive of a malignant change on microscopy, in our patient. Another interesting finding in our patient was the foul smelling serosanguinous discharge from the growth, for which two possibilities were kept in mind i.e. a secondary skin infection which was associated with allergic contact dermatitis, which was caused by the use of a local topical treatment or else, due to spontaneous necrosis of the lesion. However, absence of necrosis on microscopic examination did not support this postulation.

The histogenesis of this rare neoplasm is still unclear. A hamartomatous origin from mature apocrine, eccrine or undifferentiated pleuripotential cells has been suggested. Most lesions of syringocystadenoma papilliferum exhibit an apocrine differentiation; however, some demonstrate eccrine features. The cells which line the lumina show evidence of a decapitation secretion, thus pointing towards an apocrine origin. Results of enzyme histochemistry, immunohistochemistry and electron microscopy, obtained, have been conflicting [1].

The treatment for Syringocystadenoma papilliferum is excision biopsy, which also confirms the diagnosis [1,11]. Carbon dioxide lasers have gained importance for treatment of lesions at difficult anatomic sites, which were not suitable for surgery [12].

CONCLUSION

To conclude, solitary lesions seen in unusual locations generate multiple differential diagnoses and they must be confirmed by histopathology.

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