Dermoscopic Clues to Syringocystadenoma Papilliferum

Case Presentation

A 16-year-old male presented with multiple lesions on the left side of face present since birth. The lesions were asymptomatic and increasing in size since past 2–3 years. History of occasional discharge and bleeding from the lesions was present. There was no other complaint and family history was negative. General and systemic examination was within normal limits. Cutaneous examination revealed multiple grouped red to brown, dome-shaped, firm nodules and plaques with erosions and crusting over the surface present on the left pre-auricular area. Few lesions had central umbilication [Figure 1]. Dermoscopy done on an umbilicated nodule (marked with black arrow in Figure 1) using a hand-held dermoscope (DermLite 3Gen, in 10× magnification in polarized mode) revealed multiple bluish-white and reddish-white areas in background with central umbilication and polymorphic vessels. Yellowish areas representing crusting and whitish scales were also seen [Figure 2]. A 4-mm punch biopsy was performed on a representative lesion on the left side face. Histological examination revealed multiple papillary projections and cystic invaginations present in the upper dermis (H&E, 4×) [Figure 3]. On high power (H&E, 40×), papillary projections were lined by two rows of cells with inner row of high columnar cells having oval nuclei and faint eosinophilic cytoplasm and outer row having small cuboidal cells with round nuclei and scanty cytoplasm. Also, lymphoplasmacytic infiltrate was also present in the core of the papillary projections [Figure 4].

Based on history, clinical, dermoscopic, and histological examination, a diagnosis of syringocystadenoma papilliferum (SCAP) was made and the patient was referred to a plastic surgeon for excision of lesion.

Discussion

SCAP is a rare, benign, hamartomatous, adnexal tumor which arises from pluripotent cells exhibiting eccrine or apocrine lineage. Apocrine origin is more favored. About 50% of the cases are present at birth and up to 15–30% of the cases are seen at puberty.[1] The most frequent location is the head and neck, with scalp being the preferred location. It may present as three common types: plaque type, solitary nodular type, and linear type. Plaque type is commonly present on scalp which enlarges during puberty to become nodular, verrucous, or crusted. Linear type presents as multiple reddish pink firm papules or umbilicated nodules commonly occurring over face and neck. Solitary nodular type, in contrast, presents as nodules with predilection for the trunk, shoulder, and axillae.[2] The present case is a combination of plaque and linear type of SCAP.

SCAP is known to be associated with other benign neoplasms such as sebaceous nevus of Jadassohn, apocrine adenoma, hidradenoma papilliferum, trichoblastoma, and others.[3] Association of SCAP with basal cell carcinoma is seen in up to 10% of the cases. Histopathologically, it presents as irregular papillary projections of scaly epithelium, forming ductile structures, aligned by glandular epithelium formed by an external layer of cuboid cells, with round nuclei and scarce cytoplasm, and an internal layer of cylindrical cells with decapitation secretion and plasmacyte-rich inflammatory infiltrate.[4] Tumor cells stain positive for carcinoembryonic antigen. Dermoscopic findings in SCAP include milky red papillomatous projections with a central ulceration or crater. Polymorphic vessels can be appreciated within the darker ulcerated areas.[5] We noted multiple, bluish-white to reddish-white, structureless areas with central umbilication and polymorphic vessels. Whitish scaling and yellowish crusting were also noted in our patient. The central crater noted on dermoscopy corresponded to the characteristic cystic invagination in histopathology, whereas the white structureless area represented the surrounding hyperplastic epidermis.[6]

Dermoscopy can help play an important role in clinching the diagnosis. The dermoscopic findings of syringocystadenoma papilliferum and its differentials are summarized in Table 1.[5,6] Treatment options of SCAP include surgical excision or ablation with CO_2 laser.

In conclusion, SCAP is an uncommon tumor in the head and neck area in the pediatric age group, which may rarely undergo malignant transformation. Early diagnosis and removal of the lesion are crucial.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

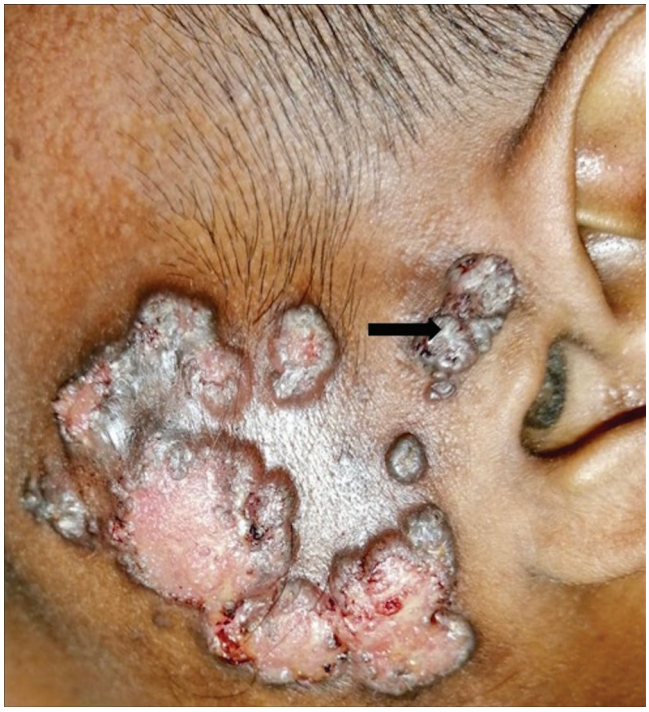

Figure 1: Multiple grouped red to brown, dome-shaped, firm nodules and plaques with erosions and crusting over the surface present on the left pre-auricular area. Few lesions had central umbilication. Black arrow marks area where dermoscopy was performed

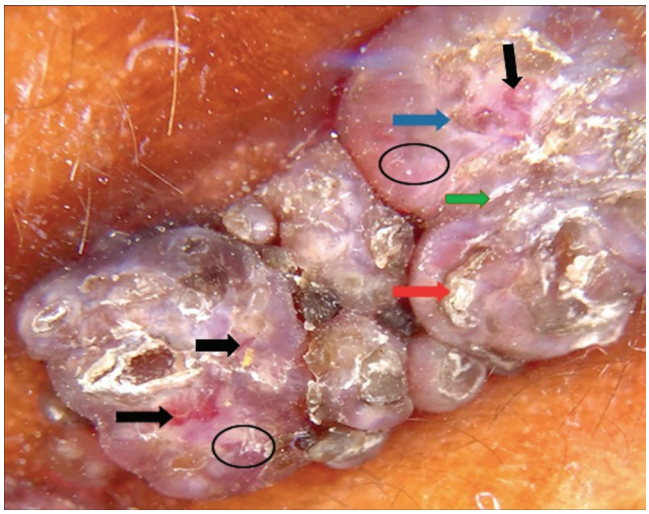

Figure 2: Multiple bluish-white and reddish-white areas in background (black circles) with central umbilication (blue arrow) and polymorphic vessels (black arrow). Yellowish areas representing crusting (red arrow) and whitish scales (green arrow) were present (DermLite 3Gen, 10×, polarized mode)

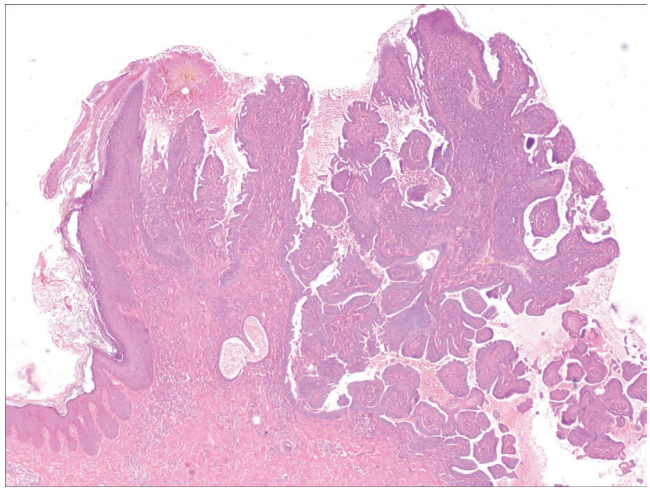

Figure 3: Multiple papillary projections and cystic invaginations present in the upper dermis (H&E, 4×)

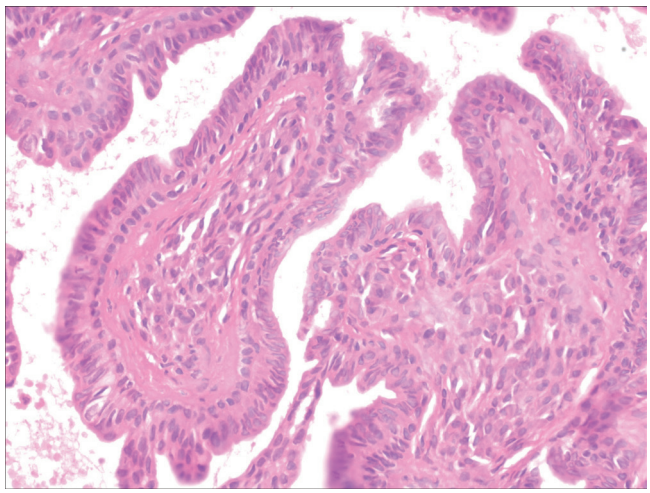

Figure 4: Papillary projections in the upper dermis lined by two rows of cells with inner row of high columnar cells having oval nuclei and faint eosinophilic cytoplasm and outer row having small cuboidal cells with round nuclei and scanty cytoplasm. Mild lymphoplasmacytic infiltrate was present in the core of papillary projections (H&E, 40×)

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Conflicts of interest

There are no conflicts of interest.

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Table 1: Dermoscopic features of SCAP and its differential diagnosis

Diseases	Dermoscopic features
Apocrine hidrocystoma	A homogeneous area that occupies the whole lesion with arborizing vessels.[5]
Tubular apocrine adenoma	Coexistence of short fine telangiectasias and large blue-gray ovoid nests arranged in a floriform pattern.[5]
Molluscum contagiosum	Coexistence of orifices, polylobular yellowish-white material, or uniform homogeneous color in the center, vascular structures, and white rosettes. This vascular pattern is restricted to the amorphous structure and never crosses the core in contrast to syringocystadenoma papilliferum.[6]
Linear unilateral basaloid follicular hamartoma	Brown-gray globules and dots, in focus dots, brown linear and arciform structures, crown vessels, short fine telangiectasias, spoke wheel-like structures without a central dark point, white structureless areas with telangiectasias, and keratotic plug.[5]
Syringocystadenoma papilliferum	Milky red papillomatous projections with a central ulceration. White circles can be seen over the rim of the milky red areas. Polymorphic vessels can be appreciated within the darker ulcerated areas.[5]

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