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Syringocystadenocarcinoma papilliferum at unusual site: inherent lesional histologic polymorphism is the pathognomon

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DESCRIPTION

Syringocystadenocarcinoma papilliferum is an extremely rare cutaneous adnexal carcinoma, considered by the WHO as the malignant form of syringocystadenoma papilliferum (SCAP).¹

The rarity of this tumour has been referred to, in a recent paper² which reviews the entity and lists a total of 12 reported cases till 2010, since it was first described in 1980.³ Eight of these 12 cases were reported to have occurred in the head and neck region (six on the scalp, one in the neck, one on the auricle), sites other than head and neck were distinctly uncommon.²

This malignant tumour having a predilection for the scalp, in older people, is a composite of a SCAP-like lesion superficially and an infiltrative adenocarcinoma in the deeper component.⁴

The benign precursor usually antedates the malignancy by many years, as a non-progressive verrucous lesion;

and when congenital, there is a frequent association with naevus sebaceous.

In older people, the presence of solitary cutaneous adenocarcinoma leads to clinical consideration of a metastatic versus primary cancer; systematic morphologic analysis, will usually resolve the issue, as illustrated in the present tumour, which occurred in a 62-year-old male, in the axilla, where syringocystadenocarcinoma papilliferum is practically unknown. As described by the patient, a bead like, 1.0 cm axillary swelling, had been self-discovered and this lesion had excoriated and attained a size of 3.5 cm in 6 months. The clinical differentials included malignant change in a pre-existing adnexal neoplasm and metastatic malignancy in a superficially located axillary lymph node.

The histology showed a villiform superficial portion, in continuity with 'ducts' bearing the 'in situ' component of the tumour and the deep, infiltrating adenocarcinoma –

Figure 1 Composite micrograph showing the papillary superficial portion of the tumour (1, 2, 4), the 'in situ' component (1, 4); and deeper located infiltrating adenocarcinoma (1, 2, 3).

Figure 2 Composite micrograph depicting the morphologic heterogeneity – papillary intraductal pattern with decapitation secretions(3), clinging (1), cribriform (2) and columnar cell (4), patterns.

distinguished by acini, tubules and ducts, with polymorphous architecture (figure 1).¹

Cribriform, papillary intraductal, clinging and columnar cell patterns with focal apocrine like decapitation secretions observed, closely resembled the similarly named mammary counterparts figure 2.^{4 5}

The educational message highlighted is that syringocystadenocarcinoma papilliferum, will very rarely occur at atypical locations and the distinctive histology should afford ineffable means of an accurate diagnosis.

Competing interests None.

Patient consent Obtained.

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