Quiz

An asymptomatic swelling on the neck

A 24-year-old male presented to our clinic for evaluation of a single, asymptomatic swelling present on the posterior aspect of his neck for the last six months. The swelling was initially small, approximately around 0.5 cm, but gradually increased in size over a period of 6 months. There was a history of a similar lesion at the same site 1 year ago, which was excised under local anesthesia.

Cutaneous examination revealed a non-compressible cystic swelling of 3 cm × 4 cm size on the left side of the nape of neck [Figure 1a]. The overlying skin had an erythematous hue. [Figure 1b].The transillumination test was positive [Figure 1c]. Within the cystic swelling, a solid, non-tender, firm mass of approximately 2 × 3 cm was palpated.

A high-resolution ultrasonography of the lesion showed a well-defined hypoechoic round mass lesion in the — subcutaneous tissue with a hyperechoic center and specks of calcification [Figure 2a]. A color Doppler study showed increased vascularity in the lesion and a branching tree pattern of blood vessels [Figure 2b].

On histopathological examination, the tumor mass was seen separate from the dermis and did not have a cellular lining. The solid tumor mass showed basophilic nucleated cells in the periphery and anucleated cells with pink keratinized cytoplasm in the center, showing abrupt keratinization with foci of calcification [Figure 3a–d].

WHAT IS YOUR DIAGNOSIS?

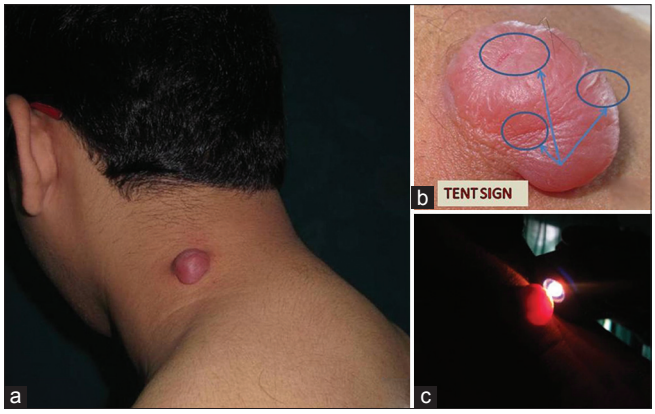

Figure 1: (a) Translucent, non-compressible cystic swelling of size 3 × 4 cm on the left side of the nape of neck. (b) The overlying skin had an erythematous hue with normal skin markings. The surface of the tumor showed multiple facets and angles (c) Transillumination test was positive

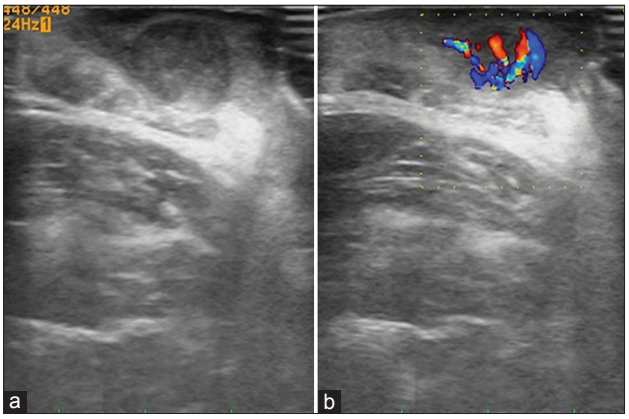

Figure 2: (a) High-resolution ultrasonography of the lesion showed a well-defined hypoechoic round mass lesion in the subcutaneous tissue with hyperechoic center and specks of calcification. (b) Color Doppler study showed increased vascularity in the lesion with branching tree pattern of blood vessels

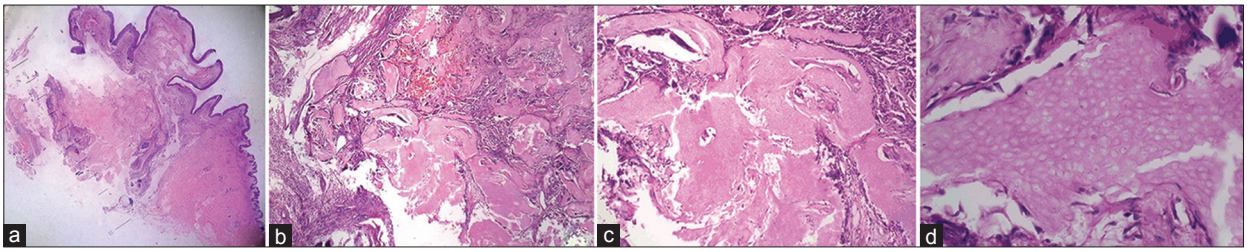

Figure 3: (a) The tumor mass was separated from dermis without any cellular lining. There was evidence of inflammatory infiltrate surrounding the tumor. (H and E, ×40) (b and c) The solid tumor mass showed basophilic nucleated cells in the periphery and anucleated cells with pink keratinized cytoplasm in the center, showing abrupt keratinization with foci of calcification (H and E, ×100). (d) Higher magnification showed basophilic nucleated cells in the periphery and anucleated cells with pink keratinized cytoplasm in the center (H and E, ×400)

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ANSWER

Cystic pilomatricoma

Pilomatricoma is an appendegeal tumor which was first described by Malherbe and Chenatais in1880 as "calcified epithelioma of Malherbe,"[1] while Dubreuilh and Cazenave described its unique histopathology consisting of basaloid cells and shadow or ghost cells in 1992.[2] Turhan and Krainer determined the origin of this neoplasm from hair cortex cells and subsequently, Forbis and Helwig renamed this neoplasm as "pilomatrixoma" which was further changed to "pilomatricoma" in 1977.[3,4]

It represents 0.12% of all skin tumors. Pilomatricomas are more common in the first and second decades of life with 40% occurring before 10 years of age and 20% occurring before 20 years. Some authors have reported a bimodal peak during the first and sixth decades of life.[5]

The tumour is more common in females with a female:male ratio of 1.5 to 2.5:1.[6] The sites of predilection include the head and neck (68.2%), followed by the trunk (14.4%), upper (15.3%) and lower (2%) extremity in decreasing order of frequency. The reported associations of pilomatricoma include myotonic dystrophy, Turner's syndrome, Gardner's syndrome, xeroderma pigmentosum, basal cell nevus syndrome and Sotos syndrome.

It typically presents as a slowly enlarging, irregularly shaped, nodular, non-tender mass freely movable over the subcutaneous tissue. The skin usually has a reddish to blue discoloration due to dilated blood vessels and chalky white nodules may be seen through the skin. The tent sign, i.e. multiple facets and angles on stretching of the skin over the tumor, is a pathognomonic sign for pilomatricoma. In addition, elicitation of " teeter-totter sign" (pressing on one edge of the lesion leads to protrusion of the opposite edge from the skin) and "skin crease sign" (a central longitudinal crease is elicited within a lesion when it is squeezed lightly along its margins, perpendicular to the skin tension line, using the thumbnail of each hand) help in diagnosis. Although most of these tumours have firm calcific nodules, a few of them look vascular with associated skin thinning.

Cystic pilomatricoma is a rare variant and presents a diagnostic challenge. Kaddu *et al*. proposed that the natural course of this neoplasm is a chronological process passing through four stages: (a) early: small and cystic lesions, (b) fully developed: large and cystic, (c) early regressive: foci of basaloid cells, shadow cells, and lymphocytic infiltrate with multinucleated giant cells without an apparent epithelial lining, and (d) late regressive: numerous shadow cells, absence of basaloid and inflammatory cells, calcification and ossification may be present. The lesion presents as an infundibular matrix cyst at an early stage, and the rapid proliferation of matrix and supramatrix cells in fully developed tumors results in an increase in the size of the lesion with time. This is accompanied by obliteration of the original cystic configuration so that they are cystic neoplasms rather than true cysts due to the absence of a cyst lining.[7] This progressive degeneration suggests episodes of microvascular bleeding that may have led to cyst formation.[8]

Based on these criteria, the tumor in our case was probably in the early regressive stage and hence presented as an atypical cystic lesion.

Ultrasound studies show a combination of hypoechogenicity, heterogenicity, and internal calcification of scattered-dot pattern. A hypoechoic rim helps in the diagnosis of pilomatricoma and in differentiating it from other subcutaneous tumors.[9] Peripheral vascularity indicated by Doppler flow signals is seen in 70% of these tumors, whereas central and peripheral vascularity is seen in only 10% of tumors (as seen in our case).[10]

Complete surgical excision is the treatment of choice. Recurrence after surgery is rare, with an incidence of 0–3%. Pilomatrix carcinomas have been observed in patients older than 40 years of age, with a tendency to local invasion and distant metastases. Malignant transformation in pilomatricoma is rare, with only 90 cases reported in literature and should be suspected in cases with repeated local recurrences. Recurrences may be related to incomplete resections.

In summary, there have been several case reports in the literature describing the clinical features and variants of pilomatricoma yet the diagnosis of atypical variants like cystic pilomatricoma continues to pose a challenge.

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