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LETTER - CLINICAL

Hybrid tumor 'spiradenocylindroma' with unusual dermoscopic features☆

Dear Editor,

Spiradenocylindroma is an adnexal neoplasm that shows histopathological characteristics of both spiradenoma and cylindroma.[1] Cylindromas and the hybrid tumor 'spiradenocylindromas' are most commonly located on the face and scalp.[1] Dermoscopy may provide significant clues for the diagnosis of adnexal tumors. Herein, we would like to report a rare case of spiradenocylindroma with distinctive dermoscopic features.

A 78-year-old woman with a history of hypertension was appointed to our clinic with the complaint of a nodule on the right forehead present for eight years. A detailed history taken from the patient revealed that the nodule was excised seven years ago but recurred within the last year. Dermatologic examination revealed a pinkish, shiny, telangiectatic nodule on the right forehead (Fig. 1). In order to aid with the diagnosis, a dermoscopy was performed which revealed thick arborizing vessels, a blue-white veil, and shiny white streaks and dots/clods resembling rosettes on a violaceous-milky red background (Fig. 1). Our differential diagnoses were nodular basal cell carcinoma, pilomatrixoma, sebaceous carcinoma and dermatofibrosarcoma protuberans. A punch biopsy was performed from the nodule and showed lobules surrounded by and containing eosinophilic basement membrane-like material. The islands were formed by two types of cells of which the peripheral ones are darker with less cytoplasm and the central ones are paler with vesicular nuclei. Spiradenomatous parts were characterized by small basaloid cells with intraepithelial lymphocytes and lymphatic like material in the stroma The two types of elements were intermingled (Figs. 2 and 3). Immunohistochemical staining features are shown in Fig. 4. The final diagnosis was spiradenocylindroma and total excision was performed.

Spiradenocylindromas are benign cutaneous adnexal neoplasms that are derived from apocrine or eccrine glands, demonstrating histopathological features of both spiradenoma and cylindroma.[1] Since a variety of benign and malignant skin disorders such as basal cell carcinoma, dermatofibrosarcoma protuberans, trichoepithelioma, and microcystic adnexal carcinoma may be considered in the differential diagnoses, histopathological examination remains the gold standard. Spiradenocylindromas are characterized by basaloid cells with eosinophilic basement membrane and tubular structures forming a multinodular pattern as the spiradenomatous part; whereas the cylindromatous portion is mainly composed of small nests of triangular or polyhedral tumoral cells forming a complex pattern resembling a jigsaw puzzle.[2] Dermoscopy may provide helpful clues to aid in the diagnosis. Senarega et al.[3] reported a case of spiradenocylindroma which showed linear vessels on a pinkish accompanied by homogeneous blue pigmentation at the periphery. Interestingly, our case showed shiny white streaks along with closely aggregated bright white clods resembling four-dots, five-dots, or cross-like rosettes.[4] Even though rosettes are most characteristically associated with actinic keratosis, and squamous cell carcinoma; they may also be seen in basal cell carcinoma, melanocytic nevus and dermatofibroma.[5] Rosettes result from the crossed polariza-

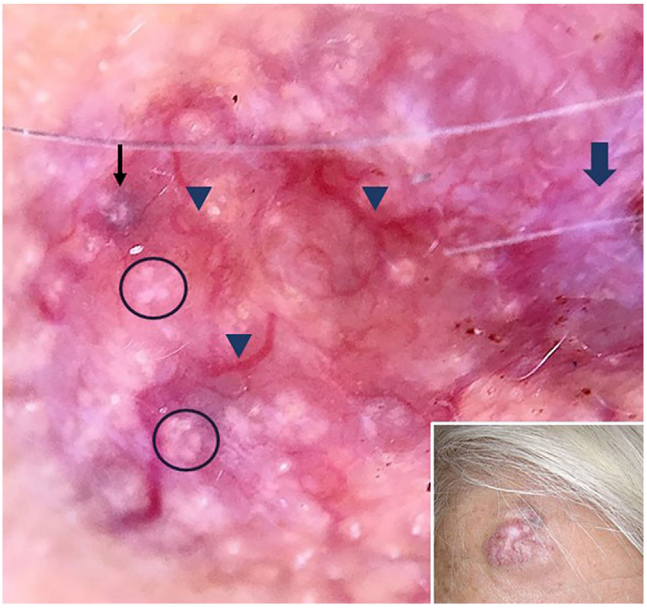

Figure 1 Pink shiny nodule on the right forehead (inset), polarized dermoscopy showed blue/gray-white veil (thin black arrow), bright-white clods and dots resembling rosettes (circles), arborizing vessels (arrowheads) and white shiny streaks (thick arrow).

☆ Study conducted at the Department of Plastic and Reconstructive Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey.

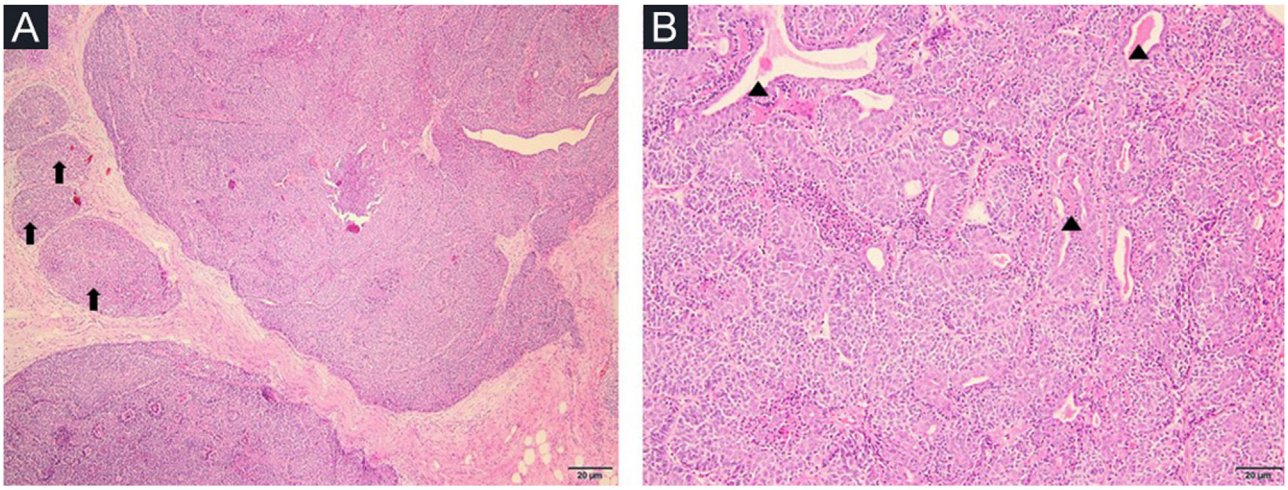

Figure 2 Nodules of different sizes (arrows); larger ones representing mostly the spiradenomatous component (A) (Hematoxylin & eosin, ×40). Bilayered ductal elements within solid aggregations, intraluminal secretory material (arrowheads) (B) (Hematoxylin & eosin, ×100).

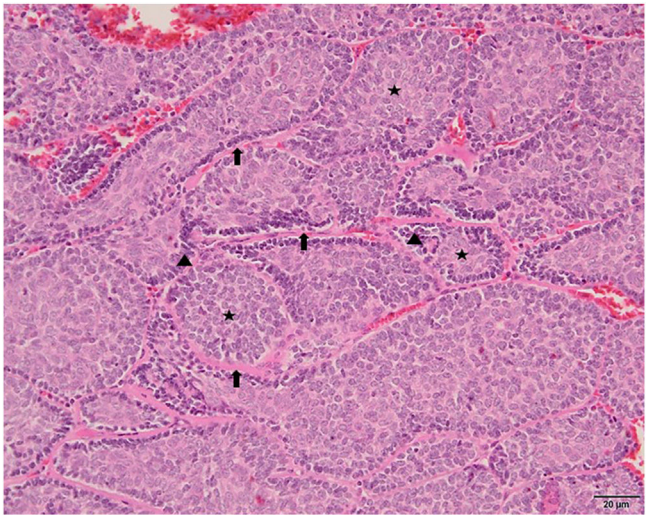

Figure 3 Jigsaw puzzle-like arrangement of islands of cells surrounded incompletely by eosinophilic basement membrane-like material (arrows). Peripheral basophilic palisading cells (arrowheads) with centrally located paler elements (asterisks) (Hematoxylin & eosin, ×200).

tion of horny material in adxenal structures or perifollicular fibrosis.[5]

To our knowledge, our patient is the first spiradenocylindroma case that shows rosette-like structures dermoscopically. By defining dermoscopic features of cylindroma, we want to emphasize the fact that spiradenocylindromas can demonstrate dermoscopic findings such as blue-gray clods and rosette-like structures which may also be detected in other cutaneous neoplasms leading to diagnostic confusion.[5]

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Authors' contributions

Ecem Bostan: Preparation and writing of the manuscript; Data collection, analysis and interpretation; Critical literature review.

Etkin Boynuyogun: Data collection; Approval of the final version of the manuscript.

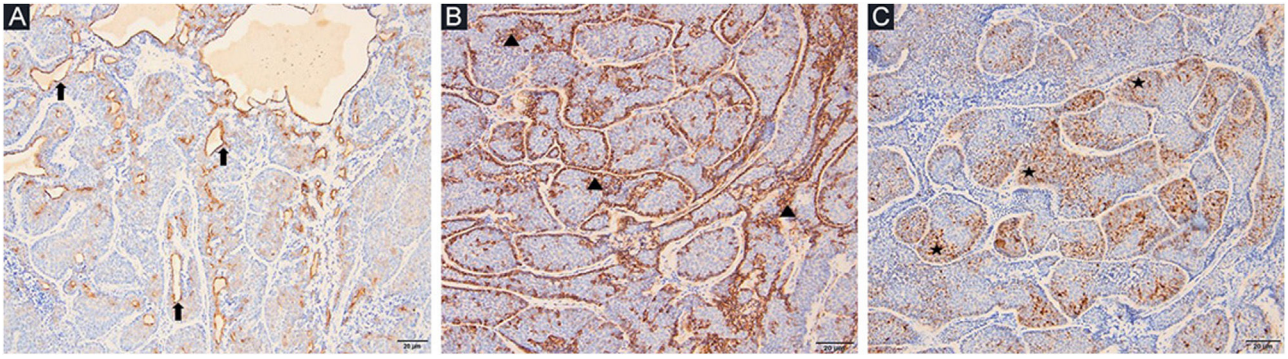

Figure 4 CK5/6 positivity in areas with ductal differentiation (arrows) (A) (CK5/6 ×100). Peripheral layer of myoepithelial cells (arrowheads) (B) (smooth muscle actin ×100). Intratumoral dendritic cells (asteriks) (C) (S100 ×100).

Ozay Gokoz: Data collection, analysis and interpretation; Approval of the final version of the manuscript.

Ibrahim Vargel: Data collection, analysis and interpretation; Approval of the final version of the manuscript.

Conflicts of interest

None declared.

References

1. Baliyan A, Dhingra H, Kumar M. Spiradenocylindroma of skin: a hybrid tumor. Indian J Dermatopathol Diagn Dermatol. 2018;5:66–8.
2. Michal M, Lamovec J, Mukensnabl P, Pizinger K. Spiradenocylindromas of the skin: tumors with morphological features of spiradenoma and cylindroma in the same lesion: report of 12 cases. Pathol Int. 1999;49:419–25.
3. Senarega A, Flores L, Innocenti AC, Parra V. Dermoscopic Features of Spiradenocylindroma. Actas Dermosifiliogr. 2019;110:604–6.
4. Lozano-Masdemont B, Polimón-Olabarrieta I, Marinero-Escobedo S, Gutiérrez-Pecharromán A, Rodríguez-Lomba E. Rosettes in actinic keratosis and squamous cell carcinoma: distribution, association to other dermoscopic signs and description of the rosette pattern. J Eur Acad Dermatol Venereol. 2018;32:48–52.
5. Liebman TN, Rabinovitz HS, Dusza SW, Marghoob AA. White shiny structures: dermoscopic features revealed under polarized light. J Eur Acad Dermatol Venereol. 2012;26:1493–7.

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Case for diagnosis. Vascular malformations, hemihypertrophy and macrodactyly: Proteus syndrome☆

Dear Editor,

A one-year-old boy had erythematous-violaceous macules on the left lower limb and trunk since birth (Fig. 1), associated with feet and chest deformities (Figs. 2 and 3), arteriovenous fistulas, and hypospadia. His personal and family history showed normal delivery at term, with no complications and non-consanguineous parents, with no reports of similar cases in the family, or hereditary diseases.

The investigation showed a normocephalic child, cervicothoracic scoliosis, posteriorly rotated ears, straight palpebral fissures, enlarged nasal base, retrognathia, flattened nasal philtrum, high palate, downturned oral commissures, and a palpable mass in the right epigastric region. He also had hemihypertrophy of limbs, enlarged hands and toes (symmetrically), and increased feet volume (left foot larger than the right one) with syndactyly between the second and the third and between the fourth and the fifth toes on the right. Vascular malformations were observed in the left lower limb, dorsum, thorax and genital region, besides linear epidermal nevus on the thorax. He had adequate neuropsychomotor development, without ocular alterations. The genetic analysis disclosed a male karyotype (46, XY), with no qualitative or structural alterations.

What is your diagnosis?

a Proteus syndrome
b Maffucci Syndrome
c Klippel-Trenaunay-Weber syndrome
d Milroy Disease

Discussion

Named in 1983 by Wiedmann et al.,[1] Proteus syndrome is characterized by its polymorphism, variable phenotypic presentations, and mosaic distribution of lesions.[2] It presents immediately at birth and can affect any organ or system, commonly manifesting with skeletal malformations, overgrowth of connective and muscular tissues, nevi and vascular malformations. Neuropsychological development is usually preserved.[3]

The clinical manifestations are variable, with cases ranging from focal changes (isolated macrodactyly),[4] to extensive dysmorphism that undergoes changes over time, making the diagnosis and therapeutic approach difficult.[5] It is considered a rare condition, with an average incidence of 1/10,000,000 births, and less than 150 cases reported worldwide.[6] It results from a mosaic mutation with somatic activation of the AKT1 oncogene (14q32.3), which is involved in cell-growth signaling pathways,[7] in addition to being associated with a greater predisposition to neoplasms, deep vein thrombosis, and pulmonary embolism, with a risk of early death.[8]

☆ Study conducted at the Department of Infectology, Dermatology, Diagnostic Imaging and Radiotherapy, Faculty of Medicine, Universidade Estadual Paulista, Botucatu, SP, Brazil.