

A row of nodules on the right side of the face



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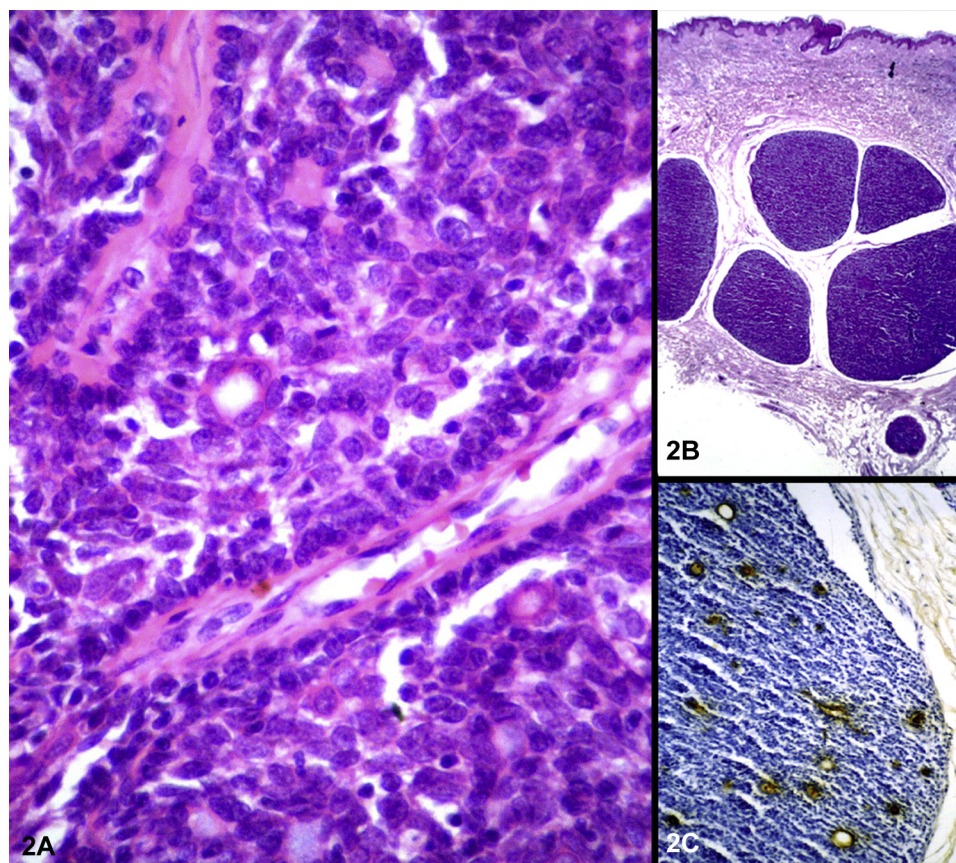
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A 38-year-old woman attended the consultation because of a row of reddish-brown papules with superficial telangiectasias on the right side of her face (Fig 1). The lesions were painful to palpation and arranged in a curved line from the inner canthus to the labial commissure. Although her parents had observed the papules since her childhood and interpreted them as nevi, the patient was now concerned about the appearance of new satellite lesions. A punch biopsy taken from a papule was stained with hematoxylin and eosin (Fig 2, A, original magnification: $\times 400$; B, original magnification: $\times 100$) and immunohistochemistry using antibodies against carcinoembryonic antigen (Fig 2, C, original magnification: $\times 250$).

Question 1: Considering the clinical and histopathologic images, what is the most likely diagnosis?

- A. Syringomas
- B. Angiofibromas
- C. Trichoepitheliomas
- D. Spiradenomas
- E. Eccrine nevus

Answers:

A. Syringomas — Incorrect. Syringomas present as small, skin-colored papules in the periocular area. The lesions tend to appear in adolescence; they are not typically congenital.

B. Angiofibromas — Incorrect. Angiofibromas are usually numerous and can be found on both sides of the face in patients with tuberous sclerosis. There are also special types of angiofibroma that are not associated with genetic syndromes, including fibrous papules in the nose and pearly penile papules. Histology would show a fibrovascular proliferation.

C. Trichoepitheliomas — Incorrect. Trichoepitheliomas are small, multiple, bilateral lesions that appear after puberty and increase in numbers with age. Histology shows rudimentary hair follicles.

D. Spiradenomas — Correct. Spiradenomas are skin-colored, pink or red papular lesions of elastic consistency that most commonly appear on the scalp, neck, and upper trunk. Involvement of the

face is uncommon, and even rarer are multiple tumors in a curvilinear distribution. Spiradenomas usually appear after puberty, but they can occasionally be seen in children. The appearances in this case are unusual, and the diagnosis could only be confirmed by the histologic appearance of the tumor.

E. Eccrine nevus — Incorrect. Eccrine nevi are a malformation of eccrine sweat glands. These rare lesions usually appear at birth and can produce a localized area of excessive sweating.

Question 2: Which histopathologic description corresponds with this diagnosis?

A. “Blue balls” in the dermis, unattached to the epidermis. Tumor cells, distributed in cords or islands, are of 2 types: Basaloid, or larger and pale; the latter are usually in the center of the islands, in some areas forming ductal structures. There are sparse lymphocytes infiltrating the neoplasm.

B. A well-circumscribed nodule in the dermis, composed of densely packed, interlacing bundles of elongated cells with cigar-shaped nuclei. Tumor cells are separated by small, compressed vascular channels.

C. There is a nodule occupying the dermis and subcutaneous fat, consisting of proliferated, but cytologically typical fibroblasts, surrounded by dense collagen bundles. Blood vessels are prominent and orientated perpendicularly to the skin surface.

D. “Blue balls” in the dermis, showing small horn cysts and rudimentary hair follicles, some of them reproducing hair bulbs and papillae.

E. The epidermis is normal. In the dermis, there is an increased number of dilated blood vessels and proliferated fibroblasts, surrounded by coarse collagen. The fibroblasts are often plump, elongated or stellate, and can be mononucleate or multinucleate. In some areas, a lymphocytic inflammatory infiltrate may be seen.

Answers:

A. “Blue balls” in the dermis, unattached to the epidermis. Tumor cells, distributed in cords or islands, are of 2 types: Basaloid, or larger and pale; the latter are usually in the center of the islands, in some areas forming ductal structures. There are sparse lymphocytes infiltrating the

neoplasm. — Correct. Spiradenomas are benign tumors derived from the straight intradermal eccrine duct.¹ Microscopically, they appear as nodules in the dermis and subcutaneous tissue, unconnected to the epidermis. These cellular nodules are variable in size and characteristically composed of 2 types of epithelial cells: Central, large cells with round vesicular nuclei and prominent nucleoli, and abundant pale cytoplasm, and small peripheral cells with hyperchromatic nuclei. Large cells can show positivity to carcinoembryonic antigen.

B. A well-circumscribed nodule in the dermis, composed of densely packed, interlacing bundles of elongated cells with cigar-shaped nuclei. Tumor cells are separated by small, compressed vascular channels. — Incorrect. The description corresponds to a leiomyoma. These skin tumors can be classified as piloleiomyomas, which are not encapsulated, and where the tumor cells are separated by collagen bundles, and angioleiomyomas, where the cells are separated by dilated vascular spaces. Angioleiomyomas are usually well circumscribed or encapsulated.

C. There is a nodule occupying the dermis and subcutaneous fat, consisting of proliferated, but cytologically typical fibroblasts, surrounded by dense collagen bundles. Blood vessels are prominent and orientated perpendicularly to the skin surface. — Incorrect. The description corresponds to a keloid, in which collagen bundles are thicker than those in the surrounding dermis and form deep nodules. Usually, there is also an increased amount of proteoglycans in the area.

D. “Blue balls” in the dermis, showing small horn cysts and rudimentary hair follicles, some of them reproducing hair bulbs and papillae. — Incorrect. This description corresponds to a trichoepithelioma, a hair-follicle-derived tumor, which also forms blue balls in the dermis; however, the histology of trichoepitheliomas differs markedly from this patient’s biopsy.

E. The epidermis is normal. In the dermis, there is an increased number of dilated blood vessels and proliferated fibroblasts, surrounded by coarse collagen. The fibroblasts are often plump, elongated or stellate, and can be mononucleate or multinucleate. In some areas, a lymphocytic inflammatory infiltrate may be seen. — Incorrect. This is the typical description of an angiofibroma. In cases of doubt, a denser collagen inside the lesion than in the surrounding dermis can be a useful diagnostic pointer.

Question 3: Why are these lesions aligned this way?

- A. They are aligned in a dermatomal fashion.
- B. They follow the path of facial lymphatics.
- C. They are aligned along the lines of Blaschko.
- D. They follow the clefts of Tessier.
- E. They follow pigmentary demarcation lines.

Answers:

A. They are aligned in a dermatomal fashion. — Incorrect. The lesions do not follow the dermatome of the second division of the trigeminal nerve, and it would be inaccurate to call it a “zosteriform” spiradenoma.

B. They follow the path of facial lymphatics. — Incorrect. The lymphatic drainage from the center of the face goes to the submandibular lymph nodes; it does not curve medially over the upper lip.

C. They are aligned along the lines of Blaschko. — Correct. These lesions follow the Blaschko lines on the face. These lines are believed to follow the migration of embryonic cells and do not correspond to areas served by nerves or lymphatics.² Cases of blaschkoid spiradenomas may be congenital or associated with genetic syndromes, such as incontinentia pigmenti or congenital hemidysplasia with ichthyosiform erythroderma and limb defects syndrome, or be, as in the present case, sporadic. The majority of cases of sporadic blaschkoid spiradenoma are observed in women, with an average age or presentation of 10.8 years.³

D. They follow the clefts of Tessier. — Incorrect. Clefts of Tessier are malformations that can involve skin, soft tissue, and bone. They are congenital and uncommon. Tessier classified facial clefts into 15 types, but none of them corresponds to the distribution of the lesions in this case.⁴ Besides, there were no soft-tissue or bony changes.

E. They follow pigmentary demarcation lines. — Incorrect. Pigmentary demarcation lines are physiologic, and present as a border of abrupt transition between a more pigmented skin area and a lighter surface. Malakar and Dhar⁵ observed that in the face, these lines seem to correspond to the division of the fifth cranial nerve. Pigmentary demarcation lines are bilateral.

Conflicts of interest

None disclosed.

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