Solitary desmoplastic trichoepithelioma is a benign adnexal neoplasm first described by Brownstein and Shapiro in 1976. It can be solitary or multiple, familial, or not familial.

We present the case of a young man affected by solitary nonfamilial desmoplastic trichoepithelioma of the external auditory canal (EAC). Clinical and histologic findings are described. To our knowledge, this is the first reported case of solitary desmoplastic trichoepithelioma of the EAC.

CASE REPORT

A 19-year-old man presented with a 2-year history of a slowly enlarging, asymptomatic lesion localized in the anterior wall of the right cartilaginous EAC. Physical examination showed a round swelling measuring approximately 2 cm in diameter, with slightly irregular outlines. It was smooth, hard, and fixed to the superficial planes but not to the deep planes. The overlying skin was normal.

Magnetic resonance imaging (MRI) showed a lesion with slightly irregular borders. The mass appeared hypointense on T1-weighted images and hyperintense on T2-weighted sequences and showed homogeneous enhancement after gadolinium infusion without signs of infiltration of the adjacent structures of the EAC.

Surgery was planned, and the lesion was removed completely from the external auditory canal. Intraoperatively, it seemed to be adherent to the tragus cartilage and epidermis so that it was necessary to remove parts of the cartilage and overlying skin to avoid the risk of leaving residual disease.

Histologic examination showed in the dermis a tumor composed of islands of basaloid cells with scarce cytoplasm set in a desmoplastic stroma and small keratinous cysts (Figs. 1–3). These findings were consistent with a diagnosis of desmoplastic trichoepithelioma (DTE). At follow-up, 1 year after surgery, the patient was tumor free.

DISCUSSION

DTE is a benign cutaneous neoplasm of adnexal origin with an incidence of 2 per 10,000. It was first described by Brownstein and Shapiro in 1976 (1), and it originates from a hair germ. A predilection for young and middle-aged female patients has been observed (1–3).

Hair germ tumors include a variety of pathologies with varying degrees of follicular differentiation, ranging from highly differentiated tumors (i.e., hair follicle nevus, trichofolliculoma) to tumors with limited epithelial differentiation. In this latter group are included the rare trichoblastic tumors. They include variants, such as the DTE, that may contain both epithelial and mesenchymal components (1,2).

FIG. 1. Islands of basaloid cells without atypia (star) in a desmoplastic stroma and small keratinous cysts (circle) (hematoxylin-eosin; original magnification, ×20).
Histologically, DTE is characterized by a triad of microscopic findings: narrow strands of tumor cells, keratinous cysts, and a desmoplastic stroma (Figs. 1–3). Tumor cells are small and basaloid with prominent oval nuclei and minimal cytoplasm, and the stroma typically contains multiple horn cysts. Mitotic figures are rare, and nuclear pleomorphism is not observed. Histologic differential diagnosis is with basal cell carcinoma, microcystic adnexal carcinoma, trichoadenoma, and trichoblastoma. Compared with these tumors, no frank cellular atypia, large tumor masses, or peripheral palisading are present in DTE (4). Because the histologic differential diagnosis may be difficult, several authors have proposed the use of immunohistochemical methods to help in the diagnosis of sclerotic epithelial neoplasm (1,5). Finally, DTEs tend to be confined to the upper two-thirds of the reticular dermis.

Clinically, it presents as a skin colored, asymptomatic, small, firm annular lesion with a raised border and a depressed center that occurs almost exclusively on the face, particularly around the nasolabial fold. In our patient, the tumor was located in the anterior wall of the EAC. To our knowledge, this is the first case of solitary desmoplastic trichoepithelioma of the EAC. It seemed to be round, firm, and mobile on the deep planes, with slightly irregular borders, exhibiting characteristics similar to a typical parotid tumor. Given its sclerotic nature and tumor strands attached to the epidermis (1), desmoplastic trichoepithelioma usually presents as a hard lesion, fixed to the superficial planes and mobile on the deep planes.

Clinical differential diagnosis is with basal cell carcinoma, sebaceous hyperplasia, granuloma annulare, scar, and scleroderma.

Therefore, an accurate imaging examination is essential to raise the suspicion of a cutaneous adnexal neoplasm. It should be emphasized that the definitive diagnosis of desmoplastic trichoepithelioma is based on the histologic examination, although clinical and radiologic considerations are useful to characterize the lesion. In our case, the signal intensity of the desmoplastic trichoepithelioma on magnetic resonance imaging examination resembled that of any benign, cutaneous mass. Desmoplastic trichoepithelioma also can present as solitary familial desmoplastic trichoepithelioma and as multiple familial and nonfamilial tumor so that an accurate evaluation of the familial history and a long-term follow-up should be carried out (1).

The treatment of choice is surgical removal through an endoaural approach. The overlying skin and other adjacent tissues must be resected to avoid recurrence.

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