EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY
Valencia, May 25-28, 2022

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Case number: M281/20
Material distributed: H&E slide

Dabska Tumor of the Eye Lid

Case Description

A 77 yo male patient presented in April 2020 with a right upper eye lid tumor. In 2018, three tumor nodules were excised elsewhere from the same area and were classified histologically as “progressive angioma”. Since 2019,recurrent progressive tumor growth was noted. On examination a firm, nodular, non-adherent lesion with vascularization, measuring 2.5 x 2 cmwas noted. The patient did not complain on double vision and or pain on eye movement. Visual acuity was 0,6 (RE) and 0,8 (LE) with incipient cataract. Further ophthalmological examinations were within normal limits. The CT scan showed a lesion of the right medial upper lid (2.9x0.6 cm) without retroseptal and/or bone infiltration. Excision of the tumor was performed on May 12, 2020.

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Macroscopy

Lid tissue. 10-26 x 4-19 x 3-5 mm needle-labeled multinodular lesion with hemorrhage

Light microscopy (H&E, PAS),Immunohistochemistry

Lid excidate with dermally situated circumscribed but not encapsulated vessel-rich lesion consisting of multiple dilated vascular structures with papillary, glomerulus-like proliferations and hobnail-cells. The cells show monomorphic enlarged hyperchromatic cell nuclei without higher-grade atypia, no mitoses and no necroses. The tumor stroma exhibits a diffuse lymphocytic concomitant infiltration. In the surrounding tissue single dilated lymphatic vessels are noted.
The tumor cells show a strong expression with CD31 and Podoplanin (D2-40), an extensive expression with ERG (Erythroblast transformation-specific (ETS) related genes) with a focal and weak expression of CD 34. No expression of Keratin and EMA. The proliferation index measured with Ki67 is only slightly elevated. The lymphocytic infiltrates demonstrate an intense expression of CD3 and CD4, only few CD8+ T lymphocytes are present.
A FISH-analysiswas negative for an amplificationof theMyc-Gen.

Diagnosis:

Papillary intralymphaticAngioendothelioma (PILA, so-calledDabska-Tumor), apparentlyarising fromlymphangioma

Differential diagnoses

- Retiform hemangioendothelioma
- Angiosarcoma
- Papillary endothelial hyperplasia (Masson’s lesion)

Discussion

A “Dabska-Tumor” was first described by Maria Dabska (Polish pathologist, 1921-2014) in 1969 as a malignant skin tumor in 6 children. She called it a “malignant endovascular papillary angioendothelioma of the skin in children”, as 2 of the 6 patients developed lymph node metastases (1). In 1999, Fanburg-Smith et al. from AFIP published 12 further cases partly in adults without recurrence and/or metastases with good long time prognosis. They called these tumors “papillary intralymphaticangioendothelioma – PILA (2). A PILA (Dabska-Tumor) typically consists of lymphatic or vascular channels lined with atypical endothelial cells and papillary endothelial proliferations surrounded by hyalinized areas (basement membrane material). Theendothelial cells are round to polyhedral with hyperchromatic apically displaced protruding nuclei and nuclear projections into the cytoplasm (hobnail cells). These hobnail cells are also found in other benign and malignant tissues eg.lactating breast, Kimura disease, nephrogenic adenoma of the bladder, adenocarcinoma of the lung, clear-cell carcinoma of the endometrium, tubulocystic renal cell carcinoma and angiosarcoma.
Lymphocytic infiltrations are typically seen in PILAs. The mitotic index is normally low. Immunohistochemistry of the tumor cells is typically positive for CD34, CD31, ERG, D2-40, and VEGFR3 (2,3). Positivity for D2-40 and VEGFR-3 may presume a lymphatic origin.
On electron microscopy, the neoplastic cells show pinocytic vesicles and so-called Weibel-Palade-bodies (4).
PILA is classified as intermediate or “borderline malignancy” – as are spindle cell, epitheloid, kaposiform, retifom and polymorph hemangioendotheliomas – and is typically assessed as angiosarcoma with low malignancy grade (5,6). Papillary intralymphaticangioendothelioma (PILA, DABSKA-Tumor) is extremely rare and generally affects skin and subcutaneous tissue of children (1,7-11). The tumor is mostly located at the head or neck area or in the extremities. It may develop in preexistent vascular malformations of the skin (12). Deeper locations eg.spleen, testes and bone were described (7,13-15). Since their first description in children, PILAs were also seen in adults with an age distribution from birth to 83 years and a mean patient age of 30 years (2).
PILA is a locally aggressive, however, rarely metastasizing tumor. A generous local excision of the tumor is the treatment of choice. In general, Dabska tumors hold a good prognosis (9,10).
As to the authors knowledge, only one further publication of a primary Dabska tumor of the eye lid in a 76 yo patient exists in the literature. This tumor also contained retiform parts and focal atypias and was finally classified as angiosarcoma. This tumor had to be excised several times for recurrences and was additionally treated with radio therapy and chemotherapy (16).

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