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Case Report

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Oropharyngeal Invasive Papillary Intralymphatic Angioendothelioma (Dabska's Tumor): About an Unusual Location of an Exceptional Tumor

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Abstract

Papillary Intralymphatic Angioendothelioma or Dabska's tumor is very rare tumor in children and young adults, characterized by intraluminal papillary endothelial structures which are lined by plump cuboidal endothelial cells with hobnailed appearance [1]. It usually develops in the soft tissues of extremities. Deeper locations are exceptional and unusual, complicating its early diagnosis and adequate management.In this paper we report a new case about a recurrent and locally invasive oropharyngeal Dabska's tumor in a 34-year-old Moroccan woman.

Keywords: Papillary Intralymphatic Angioendothelioma; Tonsil; Diagnosis pitfall

Abbreviations

PILA: Papillary Intralymphatic Angioendothelioma; PNET: Peripheric Neuro Ectodermic Tumors; WHO: World Health Organisation; CD31, CD34: Cluster Differentiation 31 and 34; ERG: Transcriptional regulator ERG; VEGFR-3: Vascular Endothelial Growth Factor Receptor-3

Introduction

Papillary Intralymphatic Angioendothelioma (PILA) or Dabska's tumor is extremely rare tumor in children and young adults Papillary. To the best of our knowledge, less than 40 PILA cases have been reported in the literature [2]. It is generally observed in the dermis and soft tissues; however, few cases were described in deeper locations such as spleen, tongue, testis and bone [1-3]. In such cases, correct diagnosis and management can be challenging.

Despite the generally indolent evolution, it can be locally invasive with the potential to metastasize [4].

Case Report

We present the case of a recurrent oropharyngeal Dabska's tumor and locally invasive in a 34-year-old woman. She was treated in 2017 for a low growing mass of the right tonsil for which multiple biopsies were performed and came back in favor of lymphoid hyperplasia then Angiosarcoma and at the end of a PNET. The patient underwent surgical excision. Three years later, clinical outcome was marked by the reappearance of the mass (Figure1).

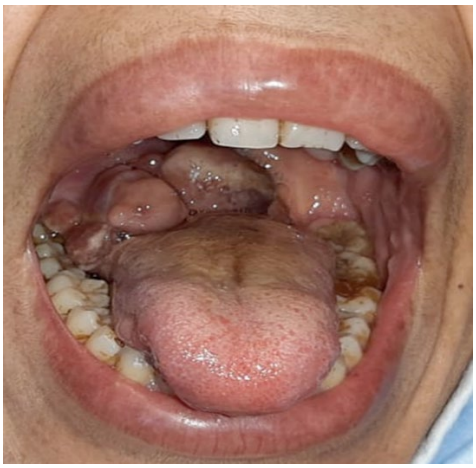

Figure 1: Clinically, we find a large, budding and ulcerated mass from the right tonsil and extending to the base of the tongue.

Computed tomography objectified a tissue process centered on the right lateral wall of the oropharynx invading the base of the tongue and the lateropharyngeal space. The patient was admitted to our institute for a new biopsy. Histologically, the tumor responded to a submucosal vascular proliferation, composed of inralymphatic clusters and anastomosing vascular channels forming papillae. These papillae had hyalinized cores and were lined by plump cuboidal endothelial cells with focal hobnailed appearance. No mitotic figures were identified (Figure2).

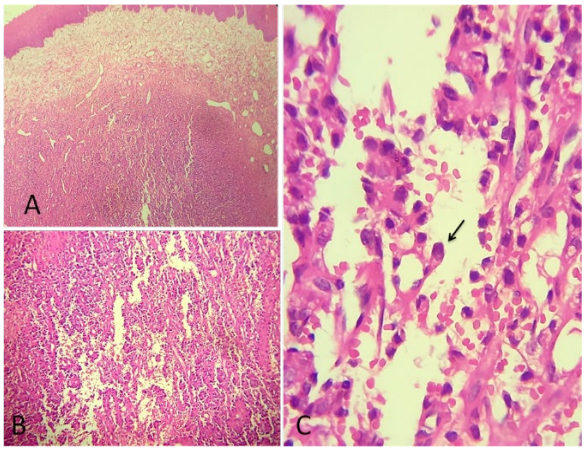

Figure 2: Histologically, (A) the tumor is composed of submucosal vascular proliferation forming papillae (Hematoxylin-eosin, original magnification x 10). (B) These papillae had hyalinized cores (Hematoxylin-eosin, original magnification x 20) and were lined by plump cuboidal endothelial cells with hobnailed appearance (C) (arrow).

Immunostaining showed a strong expression of ERG, CD31, vimentin and CD99 by tumor cells (Figure3). The Ki67 proliferation index was estimated at 4%.

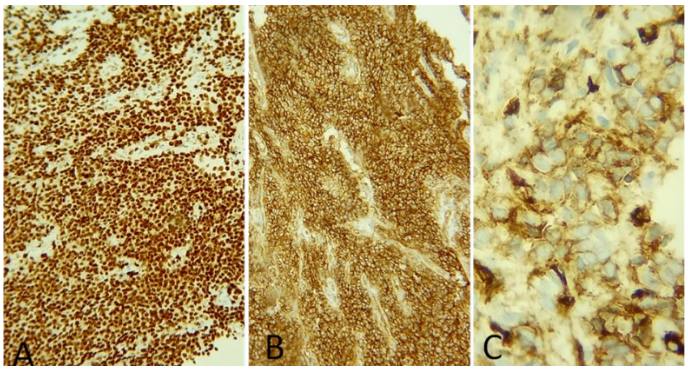

Figure 3: In immunohistochemical stain, the tumor cells expressed ERG (A), CD31 (B) and vimentin (C).

Thus, the diagnosis retained is that of an oropharyngeal Papillary Intralymphatic Angioendothelioma. Unfortunately, the patient was lost to follow-up and did not return for additional treatment.

Discussion and conclusion

Papillary intralymphatic angioendothelioma is a very rare tumor (only around forty cases reported in the literature to our knowledge) [2]. In 1969, Maria Dabska described PILA as a malignant childhood tumor in six children [1]. The tumor was described as a low-grade angiosarcoma characterized by papillary endovascular proliferations of atypical endothelial cells and anastomosing vascular channels [3]. In 2013, the latest edition of World Health Organization tumor classification (4th edition) revised the tumor description to rarely metastasizing lymphatic vascular neoplasm [3]. It is generally observed in the dermis and soft tissues; however, few cases were described in deeper locations such as spleen, tongue, testis and bone.In this paper, we report the first case (to our knowledge) of PILA occurring in the tonsil and extending to the oropharynx. Histologicaly, Dabska's tumor is characterized by intraluminal papillary endothelial structures. These papillae had hyalinized cores and were lined by plump cuboidal endothelial cells with hobnailed appearance. The cuboidal or hobnail endothelial cells lining the vascular structures are characterized by a high nuclear cytoplasmic ratio and an apically placed nucleus that produces a surface bulge, accounting for the term "hobnail" [5]. Solid areas and glomerulus-like structures may be present [1]. Mitotic figures may be seen. Many intraluminal lymphocytes may also be evident, often attached to the endothelial cells [1-5]. Immunohistochemically the tumor cells are positive for Von-Willebrand factor, CD31, CD34, ERG and vascular endothelial growth factor receptor-3 (VEGFR-3) [6].In deeper locations, it is often misdiagnosed and confused in particular with retiform hemangioendothelioma, or angiosarcoma. Retiform hemangioendothelioma and PILA share similar biologic behavior and some histologic features, the former affects mainly young and middle-aged adults with a tendency for involvement of

the limbs and trunk. The unique feature of PILA is the papillary structure lined by atypical columnar endothelial cells [7]. Ordinary angiosarcomas may also show focal morphologic patterns of PILA [7]. Angiosarcoma, however, shows more significant endothelial atypia and diffuse growth outside of blood vessels. Dabska tumor is an indolent and slowly growing tumor, but may be locally invasive and potentially metastatic [7]. Indeed, knowledge of its anatomo-clinical features and its diagnostic pitfalls - through the number of case report - would improve its prognosis and its management.

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Consent for Publication

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

Competing Interests

The authors declare that they have no competing interests.

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