

# Dabska Tumor (Endovascular Papillary Angioendothelioma) of Orbit in a Middle Aged Male: A Case Report

Dr. Liz Maria Joseph<sup>1\*</sup>, Dr. Sankar S<sup>1</sup>

<sup>1</sup>Govt Medical College, Kottayam, Kerala, India

DOI: [10.36348/sjpm.2022.v07i05.003](https://doi.org/10.36348/sjpm.2022.v07i05.003)

| Received: 02.04.2022 | Accepted: 06.05.2022 | Published: 18.05.2022

\*Corresponding author: Dr. Liz Maria Joseph  
Govt Medical College, Kottayam, Kerala, India

## Abstract

The Dabska tumor also known as Endovascular papillary angioendothelioma is a rare, low-grade angiosarcoma that often affects the skin and subcutaneous tissues of children is characterized by intraluminal papillary endothelial structures. EPA has an overall favorable prognosis. The malignant potential of this tumor should not be ignored and close follow-up should be maintained.

**Keywords:** Dabska tumor, orbit, Endovascular papillary angioendothelioma.

**Copyright © 2022 The Author(s):** This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

## INTRODUCTION

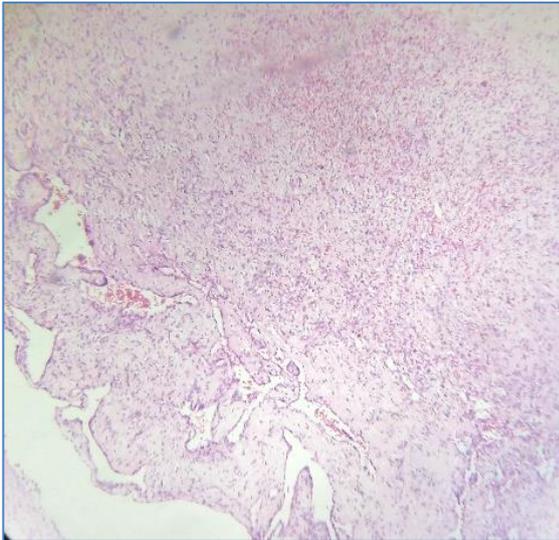
The Dabska tumor also known as Endovascular papillary angioendothelioma is a rare, low-grade angiosarcoma that often affects the skin and subcutaneous tissues of children is characterized by intraluminal papillary endothelial structures. It has a distinctive histologic architecture of anastomosing vascular channels with intravascular papillary outpouchings projecting, sometimes in a glomerulus like pattern, into a lumen lined by atypical columnar endothelial cells. In 1995, Enzinger and Weiss labelled "Dabska tumor" a form of low-grade angiosarcoma occurring in skin or subcutis of infants or young children, although we have seen rare cases in adults [1].

EPA has an overall favorable prognosis. However, it does have the potential for local recurrence and low-grade metastasis. It most frequently presents in children in various skin locations, subcutaneous tissue, and in deeper areas of the body. However, there have been reports of EPA in adults and internal organs [2]. A biopsy is diagnostic and treatment is wide surgical excision. Because of the rarity of these tumors, most of the information is available from case reports and case series [3].

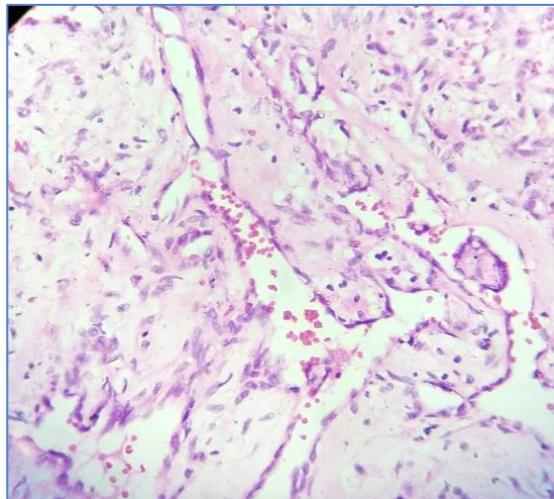
Endovascular papillary angioendothelioma is within a borderline area between benign lesions such as hemangioma and malignant ones like angiosarcoma. There have been cases of these tumors arising within preexisting vascular lesions such as cavernous hemangiomas. Morphologic similarity has also been observed with retiform hemangioendothelioma and may represent the spectrum of the same lesion. Based on the literature review, currently, the diagnosis of EPA is limited to low-grade sarcoma, demonstrating characteristic histopathological and immunohistochemistry features[4].

## CASE REPORT

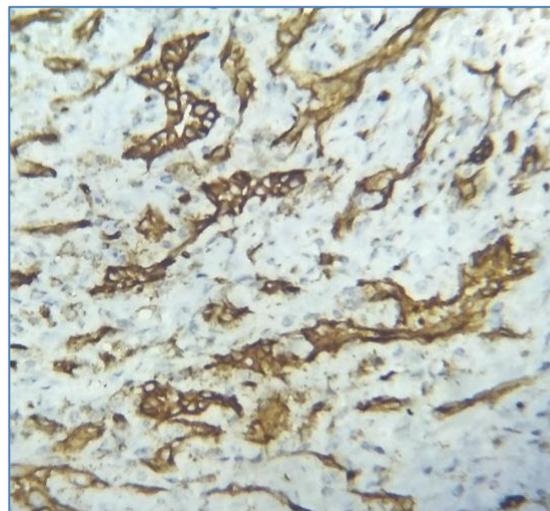
41 year old male presented with complaints of painless cystic swelling on superomedial aspect of right orbit for 2 months duration. MRI scan of orbit –Soft tissue lesion measuring 2x1x1 cm, possibly infected nasolacrimal cyst. Routine blood investigations were normal. No other comorbidities were identified. Excision of the swelling was done. Histopathological diagnosis was Endovascular papillary angioendothelioma (Dabska tumor). IHC was also done which showed CD31 and CD34 positivity.



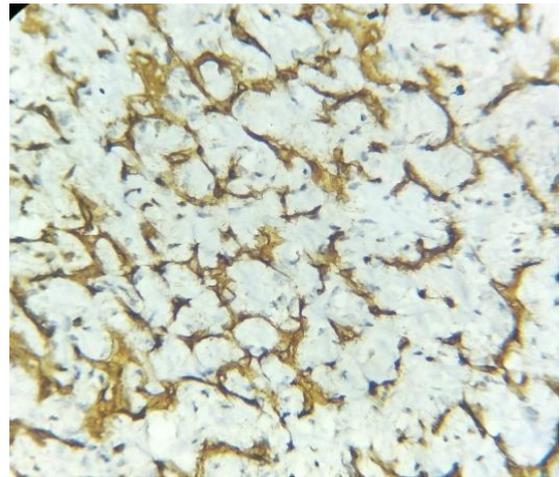
**Fig-1: Anastomosing vascular channels with intravascular papillary outpouchings projecting into a lumen**



**Fig-2: The cuboidal or hobnail endothelial cells lining the vascular structures are characterized by a high nuclear cytoplasmic ratio and an apically placed nucleus**



**Fig-3: Tumor Cells are CD31 positive**



**Fig-4: Tumor cells are CD34 positive**

## DISCUSSION

The Dabska tumor was first described in 1969 by Maria Dabska in skin and subcutaneous tissues in children and named as malignant endovascular papillary angioendothelioma. She described 6 cases in her series. Since then approximately 30 cases have been described in the literature. Of these 18 were children and 12 were adults. The age of the patients ranged from birth to 83 years and there was no sex predilection. Besides skin and subcutaneous tissue, the tumor has been described in other deeper locations like spleen, soft tissues, bone, and tongue. Generally it presents as a slow-growing, painless, usually intradermal nodule that grows to 2–3 cm in diameter[1].

The histological diagnosis of endovascular papillary angioendothelioma (EPA) (Dabska tumor) is controversial although the tumor is included in the World Health Organization classification. Papillary endothelial proliferation with a central hyaline core is one of the most characteristic features of EPA; however, this type of proliferation has been observed in other vascular tumors, such as angiosarcoma, retiform hemangioendothelioma and glomeruloid hemangioma. Several vascular tumors have EPA-like foci and EPA is not well defined generally. Endovascular papillary angioendothelioma may not be a distinct entity and may well include a heterogeneous group of lesions [1, 2].

At low power, Dabska tumor appears similar to cavernous lymphangiomias. 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" or "matchstick" [3]. Immunohistochemically the tumor cells are positive for Von-Willebrand factor, CD31, CD34 and vascular endothelial growth factor receptor-3 (VEGFR-3). The staining for CD34 and VEGFR-3 is most intense out of these [4].

Prognostically these tumors are low grade lesions with a capacity to extend to regional lymph nodes. The definitive diagnosis of endovascular papillary angioendothelioma (EPA) is made with a biopsy. As this is such a rare entity, protocols for disease involvement, surveillance and follow-up have yet to be established. It is reasonable to consider obtaining a chest radiograph for any patient with pulmonary symptoms due to a reported case of a patient dying of pulmonary metastases. Lymph node examination may be performed if there is suspicion for involvement [5]. Surgical excision is recommended. In general, the prognosis is favorable for Dabska tumors even with invasion into deeper structures. However, the malignant potential of this tumor should not be ignored, and close follow-up should be maintained. Regional lymphadenectomy should be performed when those structures appear involved [3,4].

## REFERENCES

1. Fukunaga, M. (1998). Endovascular papillary angioendothelioma (Dabska tumor). *Pathology international*, 48(10), 840-841.
2. Yamada, A., Uematsu, K., Yasoshima, H., Sakurai, K., Hori, K., Ohya, M., ... & Ogasawara, H. (1998). Endovascular papillary angioendothelioma (Dabska tumor) in an elderly woman. *Pathology international*, 48(2), 164-167.
3. Bernić, A., Novosel, I., & Križanac, Š. (2012). An unusual mole: an adult case of Dabska tumour. *Collegium antropologicum*, 36(2), 171-172.
4. Neves, R. I., Stevenson, J., Hancey, M. J., Vangelisti, G., Miraliakbari, R., Mackay, D., & Clarke, L. (2011). Endovascular papillary angioendothelioma (Dabska tumor): underrecognized malignant tumor in childhood. *Journal of pediatric surgery*, 46(1), e25-e28.
5. Eng, O. S., Borah, G. L., & Gannon, C. J. (2013). CASE REPORT Successful Treatment of a Rare Massive Dabska Tumor. *Eplasty*, 13.