INDEX COPERNICUS INTERNATIONAL

Dąbska Tumor – a rare neoplasm of the paranasal sinuses. Case report and review of world literature

Guz Dąbskiej – rzadki nowotwór zatok przynosowych. Opis przypadku i przegląd piśmiennictwa światowego

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Manuscript Preparation
E – Literature Search
F – Funds Collection

Sławomir Piotrowski[1ABDEF], Urszula Śmiałek[2B], Sylwia Kropidłowska[1CDEF]

[1]Department of Otolaryngology and Maxillary Surgery, St. Vincent de Paul in Gdynia, Poland; Head: Sławomir Piotrowski MD PhD
[2]Department of Pathology of the Gdynia Oncology Center of the Maritime Hospital Polish Red Cross in Gdynia, Poland; Head: Radosław Lenckowski MD

Article history: Received: 05.03.2021 Accepted: 22.04.2021 Published: 30.04.2021

ABSTRACT: The authors present a rare case of endovascular papillary angioendothelioma also known as "Dąbska tumor", which occurs mainly in children and young adults. This neoplasm was found in the right maxillary sinus and nasal cavity in a 22-year-old woman that remained undiagnosed for one year. After the initial diagnostic imaging, the tumor was removed through the endoscopic approach. After a year, the patient developed a mucopurulent cyst in the postoperative cavity, which was removed without any recurrence of the neoplastic disease in the follow-up histopathological examination. The authors present the clinical picture of this potentially malignant neoplasm and review the current literature.

KEYWORDS: Dabska tumor, endoscopic treatment, paranasal sinuses, rare neoplasms, sinus tumors

STRESZCZENIE: Autorzy opisują rzadki przypadek śródbłoniaka wewnątrznaczyniowego brodawkowatego, znanego również jako „guz Dąbskiej", występującego głównie u dzieci i młodych dorosłych. Nowotwór ten został stwierdzony w prawej zatoce szczękowej i w jamie nosa u 22-letniej pacjentki, u której rozwijał się niezdiagnozowany przez jeden rok. Po wstępnej diagnostyce obrazowej guz został wycięty doszczętnie na drodze endoskopowej. Po upływie roku u chorej doszło do wytworzenia torbieli śluzowo-ropnej w loży pooperacyjnej. Torbiel usunięto, a w kontrolnym badaniu histopatologicznym nie stwierdzono nawrotu choroby nowotworowej. Autorzy przedstawiają kliniczny obraz tego potencjalnie złośliwego nowotworu oraz dokonują przeglądu aktualnego piśmiennictwa.

SŁOWA KLUCZOWE: guz Dąbskiej, guzy zatok, leczenie endoskopowe, rzadkie nowotwory, zatoki przynosowe

ABBREVIATIONS

CT – computed tomography
EPA/PILA – endovascular papillary angioendothelioma /papillary intralymphatic angioendothelioma
MRI – magnetic resonance imaging

INTRODUCTION

Dąbska Tumor, or *endovascular papillary angioendothelioma* (EPA); *papillary intralymphatic angioendothelioma* (PILA) is a rare tumor in soft tissue. Most authors classify Dąbska tumor as a neoplastic entity on the border of benign lesions, such as hemangiomas, and malignant ones, such as angiosarcomas. Due to the fact that two patients had lymph node metastasis, it was initially considered a malignant neoplasm [1]. In view of its borderline behavior and pronounced lymphatic phenotype, the tumor was subsequently renamed PILA in 1998 by Fanburg-Smith et al. [2]. There have been *de novo* cases, as well as those formed within the chronic lymphedema or preexisting vascular malformation, such as, for example, hemangioma, cavernous hemangioma or peripheral lymphoma [3]. Dąbska tumors have a favorable prognosis; however, they may have the potential for local recurrence and low-grade metastases. At the time of diagnosis, lesions tend to have a size of two to three centimeters. The diagnosis of choice is biopsy, and treatment is based on a wide surgical excision [3, 4]. Because these lesions are so rare, most of the information can be found in case reports.

The authors present a description of Dąbska tumor, which was discovered in the maxillary sinus of a young woman.

 DOI: 10.5604/01.3001.0014.8653

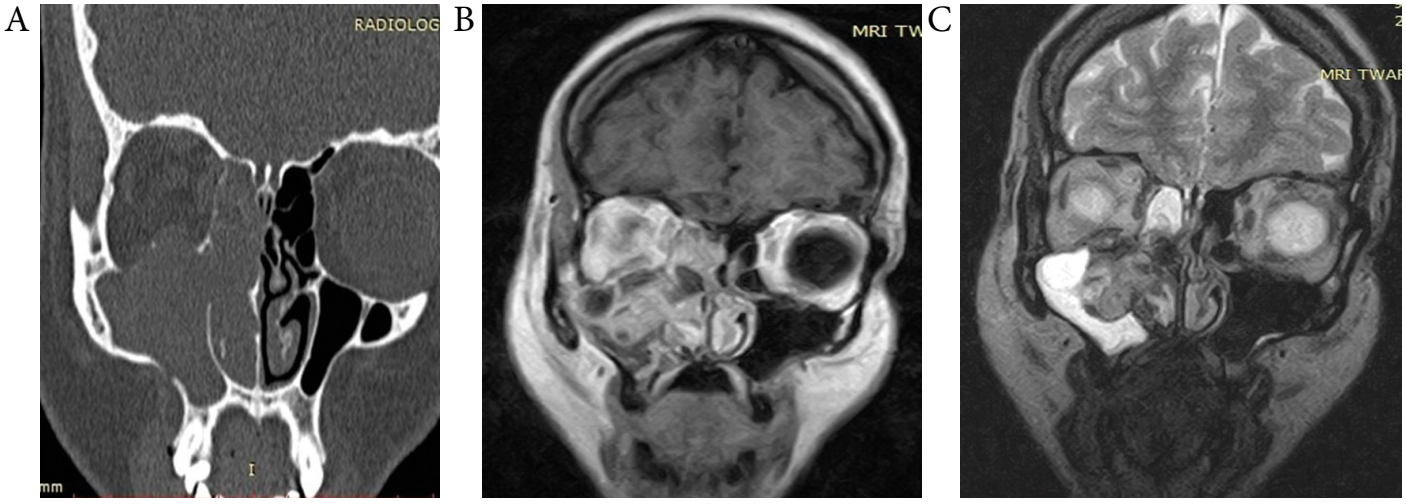

Fig. 1. (A) CT and (B, C) MRI of Dąbska tumor in right maxillary sinus. CT and MRI images show penetration of the tumor masses into the orbit and the nasal cavity. MRI clearly shows the extent of tumor and the accompanying secondary changes in the sinuses.

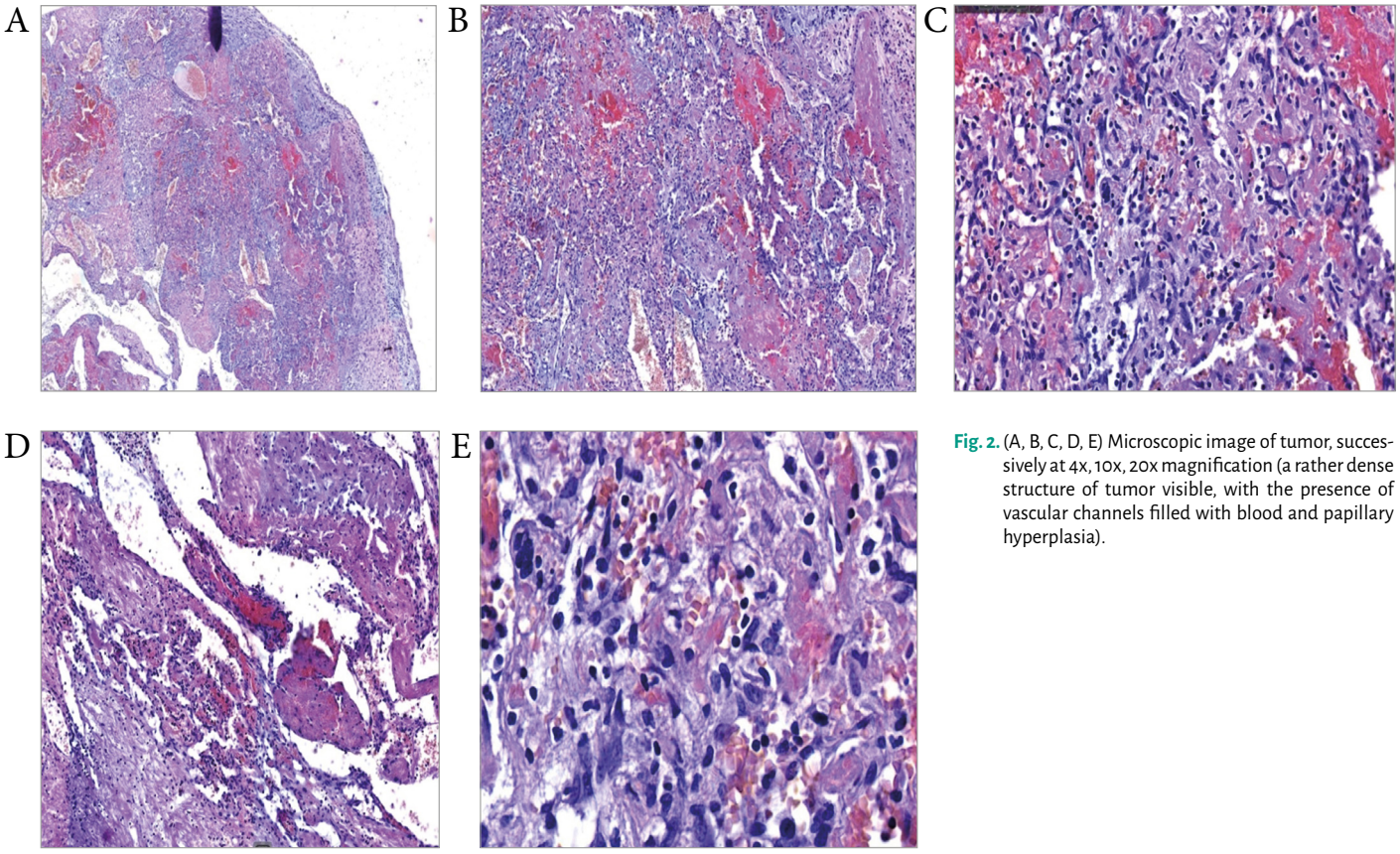

Fig. 2. (A, B, C, D, E) Microscopic image of tumor, successively at 4x, 10x, 20x magnification (a rather dense structure of tumor visible, with the presence of vascular channels filled with blood and papillary hyperplasia).

CASE REPORT

A 22-year-old female was admitted to the Department on August 30, 2020 with a tumor diagnosed in the right maxillary sinus. The patient reported: gradually worsening nasal patency on the right side, year-long recurrent secretion of yellowish nasal discharge, and bloody discharge since approximately two weeks. The patient denied any headaches or vision impairment. Physical examination revealed a polyp filling the entire nasal cavity on the right side and right-eye lacrimation. Computed tomography (CT) of the paranasal sinuses was as follows: "On the right, lesions of heterogeneous intermediate densities fill in the right maxillary sinus, the rushes and the frontal sinus, and the sphenoid sinus, as well as the middle and posterior parts of the nasal cavity, protruding approximately 6 mm beyond the posterior nasal apertures. Bone scaffold of the nasal turbinates on this side visible in sections". Magnetic resonance imaging (MRI) revealed that the main mass of the tumor is localized within the maxillary sinus, protruding into the nasal cavity, while the remaining lesions reported in the CT scan were secondary.

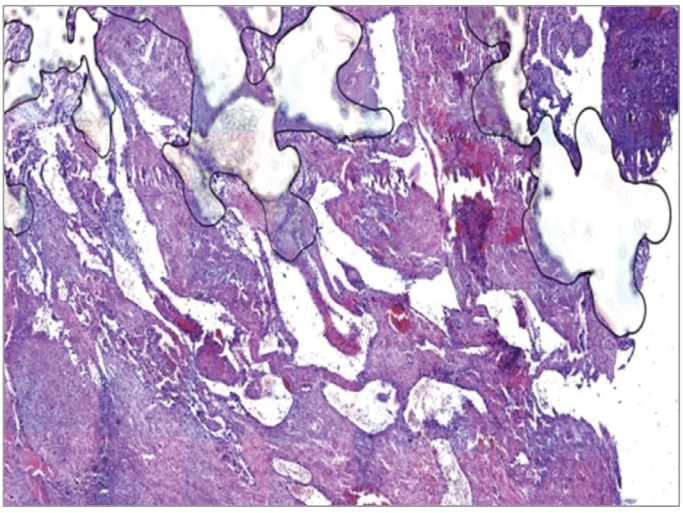

Fig. 3. Moderate atypia of endothelial cells visible at 40x magnification, as well as admixture of inflammatory cells (lymphocytes).

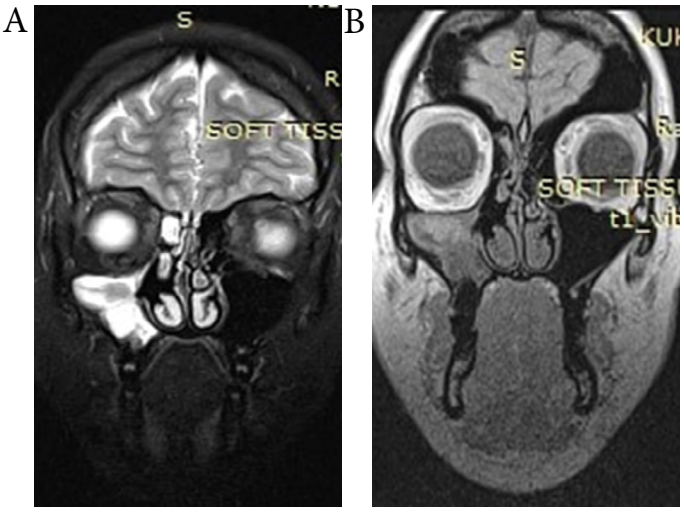

Fig. 4. (A, B) Image of follow-up postoperative MRI in the presented case. Visible cystic lesion filling the right maxillary sinus, not penetrating the eye socket nor the nasal cavity.

On August 30, 2019, right-sided endoscopic resection of the nasal and sinus tumor was performed. The tumor was accessed through the nasal cavity and via an opening made within the right canine fossa. After excision, we found numerous defects in the osseous deficiencies of the right maxillary sinus – mainly within the upper wall (visible orbital periosteum protruding under compression of the eyeball), as well as in the posterolateral portion. The procedure and further hospitalization were uneventful. The patient was discharged from the Department on the 2nd postoperative day in good general and local condition, without visual impairment. Histopathological examination of the postoperative material revealed endovascular papillary angioendothelioma (Fig. 2.).

Due to the possible malignancy of tumor, the patient required constant oncological and otolaryngological monitoring. In February 2020, the patient was diagnosed with a severe swelling of the nasal mucosa accompanied by purulent discharge – she had an ongoing upper respiratory tract infection. She was referred for a follow-up CT scan and MRI of the paranasal sinuses, however, the COVID-19 pandemic developing at that time significantly impeded diagnosis. The tests were performed in August 2020 and found: "filling of the entire lumen of the right maxillary sinus by tissue masses with a slight marginal enhancement going upwards along the wall of the orbit and into the posterior ethmoid cells (Residual lesion? mucosal lesions?)".

The patient felt well and returned for another follow-up visit in November 2020. She was qualified for re-treatment in the scope of cleaning the tumor bed and subsequent histopathological verification of the lesions. The procedure was performed on December 9, 2020. Initially, there were plans to perform only endoscopic surgery through the nasal cavity, however, due to the rather cohesive scarring changes filling the lateral wall of the nasal cavity at the entrance to the maxillary sinus, it was decided to reopen it through the fossa. This choice was additionally determined by the greater safety of this type of procedure for the soft tissues of the orbit. During the procedure, it was observed that the described lesion was probably a thickwalled mucopurulent cyst, filled with thick, purulent contents. A wide connection of the right maxillary sinus with the nasal cavity was made. The procedure and the postoperative period were uneventful. The patient was discharged on the next postoperative day in good general and local condition. The outcome of histopathological examination of postoperative material was as follows: "Sinonasal mucosa with a superimposed moderately severe chronic inflammation, no oncologically suspicious lesions". Follow-up visits revealed no changes in the nasal cavity and sinuses.

DISCUSSION

Endovascular papillary angioendothelioma was first described in 1969 in the journal "Cancer" by Professor Maria Dąbska and to this day, it is called "Dąbska's tumor" from her last name. The author presents the structure and characteristics of this tumor detected in six children aged 4 months to 6 years [1, 5]. Tumor locations included: knee, heel, cheek, temporal area, neck, hand; in three cases the tumor infiltrated deep structures such as: muscles, fascia, tendons and bones [5]. The lesion has been described as low-grade vascular sarcoma, characterized by a papillomatous intravascular growth of atypical endothelium that forms vascular channels connecting within the skin [5, 6]. Although there have been reports of regional lymph node metastases, the prognosis for long-term relapse-free survival is good [5–8]. According to Silva et al. [8] (paper from 2020), less than 40 such cases have been described so far. Some authors believe that the most frequent tumor locations are soft tissues of the skin of the extremities, although cases of involvement of deep structures such as the spleen, tongue, testicle or bones have been described. The location of Dąbska tumor within the maxilla, orbit and zygomatic bone was discussed by Li et al. [9]. A case of malignant transformation into vascular sarcoma was also presented [10]. Due to its rarity, multifocal nature and morphological features, it is a great challenge for histopathologists [8]. Looking deeper at the world literature, it turns out that it has taken professor of pathomorphology M. Dąbska 30 years of work to record the fate of 6 children diagnosed with this cancer. Two of them developed metastases. In the former case, the metastases involved the lymph nodes, while in the latter- both

Tab. I. Patients diagnosed with Dąbska tumor. The table presents 47 cases (22 children/25 adults; 25 women/22 men).

NO.	AGE	SEX	LOCATION OF LESIONS	TREATMENT	SOURCE
1	congenital change	F	knee	excision, radiation therapy	[5]
2	congenital change	M	heel	excision	[5]
3	lesion congenital	F	cheek	excision, radiotherapy	[5]
4	congenital change	M	thumb	excision	[2]
5	congenital change	F	temple	excision	[5]
6	15 months	M	facial bones (zygomatic bone, orbital bone, jaw bone)	excision	[9]
7	22 months	M	testicle	orchidectomy	[11]
8	5 years	M	spleen	splenectomy	[19]
9	6 years	M	forearm	excision	[20]
10	7 years	F [1)]	neck	excision	[5]
11	8 years	F	soft tissues of forearm	resection	[24]
12	9 years	M	shoulder blade	excision	[7]
13	11 years	F	abdomen	excision	[2]
14	approx. 11 years	F	knee	excision	[25]
15	13 years	F	palatine tonsil	tonsillektomia	[23]
16	14 years	M	thigh	-	[2]
17	14 years	M [2)]	hand	amputation	[5]
18	14 years	F	thigh	excision	[33]
19	15 years	M	thigh	-	[26]
20	16 years	F	buttock	-	[12]
21	16 years	F	buttock	-	[2]
22	18 years	M	buttock	excision	[27]
23	22 years	F	maxillary sinus	excision	current work
24	23 years	F	subcutaneous tissue of abdomen	excision	[28]
25	24 years	M	testicle	orchidectomy	[21]
26	30 years	F	arm	excision	[31]
27	32 years	F	hand	-	[2]
28	35 years	M	buttock	-	[2]
29	35 years	F	soft tissue of foot	excision	[8]
30	38 years	M	gluteus muscles	-	[2]
31	39 years	F	femur	enucleation, resection	[17]
32	41 years	M	thumb	-	[2]
33	42 years	M	vertebra	corpectomy	[22]
34	42 years	F	thigh/buttock	excision	[10]
35	43 years	M	calf	-	[2]
36	44 years	F	bone	enucleation	[18]
37	45 years	F	femur	enucleation	[18]
38	46 years	M	thigh	-	[2]

NO.	AGE	SEX	LOCATION OF LESIONS	TREATMENT	SOURCE
39	51 years	M	collarbone	excision	[12]
40	53 years	M	ear	excision	[13]
41	58 years	F	thigh	excision	[30]
42	59 years	F	abdomen	-	[2]
43	59 years	F	neck	excision	[14]
44	63 years	F	cheek	excision	[4]
45	67 years	M	tongue	excision	[29]
46	68 years	F	forearm	excision	[15]
47	83 years	F	neck	excision	[16]

[1)] metastases to the lymph nodes
[2)] metastases to the lymph nodes and lungs
F = FEMALE/M = MALE/ - = not available

the lymph nodes and the lungs, leading to death of the patient [1]. Interestingly, more recent studies often mention cases of adults, which may change the epidemiological picture of this cancer in the near future. Already in 1999, Fanburg-Smith et al. described a case series of 12 patients that covered an age interval of 8–59 years with an average of 30 years, suggesting a wider range [11].

Tab. I. contains the collected cases, including those of prof. M. Dąbska, found in available scientific works. It follows from the table that tumor incidence is not related to gender nor age. The authors did not find a description of Dąbska tumor within the paranasal sinuses in the Polish otorhinolaryngological literature, therefore they have chosen this case for presentation.

References

1. Schwartz R.A., Dabski C., Dabska M.: The Dabska tumor: a thirty-year retrospect. Dermatology., 2000; 201(1): 1–5.
2. Fanburg-Smith J.C., Michal M., Partanen T.A., Alitalo K., Miettinen M.: Papillary intralymphatic angioendothelioma (PILA): A report of twelve cases of a distinctive vascular tumor with phenotypic features of lymphatic vessels. Am J Surg Pathol, 1999; 23: 1004–1010.
3. Bhatia J., Tadi P.: Endovascular Papillary Angioendothelioma. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan. 31.
4. Bernić A., Novosel I., Krizanac S.: An unusual mole: an adult case of Dabska tumour. Coll Antropol., 2012; 36(Suppl 2): 171–172.
5. Dabska M.: Malignant endovascular papillary angioendothelioma of the skin in childhood. Clinicopathological study of 6 cases. Cancer, 1969; 24: 503–510.
6. Neves R.I., Stevenson J., Hancey M.J., Vangelisti G., Miraliakbari R. et al.: Endovascular papillary angioendothelioma (Dabska tumor): underrecognized malignant tumor in childhood. Review. J Pediatr Surg., 2011; 46(1): e25–28.
7. Morgan J., Robinson M.J., Rosen L.B., Unger H., Niven J.: Malignant endovascular papillary angioendothelioma (Dabska tumor). A case report and review of the literature. Am J Dermatopathol., 1989; 11(1): 64–68.
8. Silva T.S., deAraujo L.R., Paiva G.R., Andrade R.G..: Papillary intralymphatic angioendothelioma: Dabska tumor. An Bras Dermatol., 2020; 95(2): 214–216.
9. Li B., Li Y., Tian X., Li Z.: Unusual multifocal intraosseous papillary intralymphatic angioendothelioma (Dabska tumor) of facial bones: a case report and review of literature. Diagn Pathol., 2013; 8: 160.
10. Antosz Z., Zaniewski M., Kostecki J., Poreba R.: Angiosarcoma arising within a Malignant Endovascular Papillary Angioendothelioma (Dabska tumor) Neuro Endocrinol Lett., 2010; 31(4): 454–456.
11. Bhatia A., Nada R., Kumar Y., Menon P.: Dabska tumor (Endovascular papillary angioendothelioma) of testis: a case report with brief review of literature. Diagn Pathol, 2006; 1: 12.
12. Gambarotti M., Righi A., Sbaraglia M., Bianchi G., Picci P. et al.: Intraosseous papillary intralymphatic angioendothelioma (PILA): one new case and review of the literature. Clinical Sarcoma Research, 2018; 8: 1.
13. de Dulanto F., Armijo-Moreno M.: Malignant endovascular papillary hemangioendothelioma of the skin: The nosological situation. Acta. Derm Venereol (Stockh), 1973; 53: 403–408.
14. Miyachi Y., Imamura S.: Very low-grade angiosarcoma. Dermatologica, 1981; 162: 206–208.
15. Fukunaga M., Ushigome S., Shishikura Y., Yo-Koi K., Ishikawa E.: Endovascular papillary angioendothelioma-like tumor associated with lymphoedema. Histopathology, 1995; 27: 243–249.
16. Yamada A., Uematsu K., Yasoshima H., Sakurai K., Hori K. et al.: Endovascular papillary angioendothelioma (Dabska tumor) in an elderly woman. Pathol Int, 1998; 48: 164–167.
17. Nakayama T., Nishino M., Takasu K., Hayakawa K., Toguchida J. et al.: Endovascular papillary angioendothelioma (Dabska tumor) of bone. Orthopedics., 2004; 27: 327–328.
18. McCarthy E.F., Lietman S., Argani P., Frassica F.J.: Endovascular papillary angioendothelioma (Dabska tumor) of bone. Skeletal Radiol., 1999; 28: 100–103.
19. Katz J.A., Mahoney D.H., Shukla L.W., Smith C.W., Gresik M.V. et al.: Endovascular papillary angioendothelioma in the spleen. Pediatr Pathol, 1988; 8: 185–193.
20. Patterson K., Chandra R.S.: Malignant endovascular papillary angioendothelioma: Cutaneous borderline tumor. Arch Pathol Lab Med, 1985; 109: 671–673.
21. Anne M.: Schultheis, Mareike Sandmann, Stefan Steurer: Strong ERG Positivity in Papillary Intralymphatic Angioendothelioma of the Testis of a 24-Year-Old Male: A Case Report. Case Rep Pathol., 2013; 2013: 531479.
22. Bhat S., Kishanprasad H.L., Shetty S.M., Nirupama M., Shetty J.K.: Metastatic Carcinoma to Vertebra Mimicking as Primary Malignant Vascular Tumour; A unusual Case Report. Indian J Surg Oncol., 2016; 7(3): 341–344.
23. Mukherjee S., Mallick J., C Pal P., Chattopadhyay S.: Hemangioendothelioma of soft tissue: Cytological dilemma in two cases at unusual sites. J Cytol., 2012; 29(1): 89–91.
24. Argani P., Athanasian E.: Malignant endovascular papillary angioendothelioma (Dabska tumor) arising within a deep intramuscular hemangioma. Arch Pathol Lab Med, 1997; 121: 992–995.
25. Allen P.W., Ramakrishna B., MacCormac L.B.: The histiocytoid hemangiomas and other controversies. Pathol Annu, 1992; 27: 51–87.

26. Magnin P.H., Schrok R.G., Barquin M.A.: Endovascular papillary angioendothelioma in children. Pediatr Dermatol, 1987; 4: 332–335.

27. Quecedo E., Martinez-Escribano J.A., Febrer I., Oliver V., Velasco M. et al.: Dabska tumor developing within a preexisting vascular malformation. Am J Dermatopathol, 1996; 18: 302–307.

28. Eng O.S., Borah G.L., Gannon Ch.J.: CASE REPORT Successful Treatment of a Rare Massive Dabska Tumor. Eplasty., 2013; 13: e46.

29. Takaoka K., Sakurai K., Noguchi K., Hashitani S., Urade M.: Endovascular papillary angioendothelioma (Dabska tumor) of the tongue: report of a case. J Oral Pathol Med; 2003; 32(8): 492–495.

30. O Emanuel P., Lin R., Silver L., B Birge M., Shim H. et al.: Dabska tumor arising in lymphangioma circumscriptum. J Cutan Pathol., 2008; 35(1): 65–69.

31. Banik T., Sinha A.K., Rai M.K., Prasad J.: Fine needle aspiration cytology of malignant endovascular papillary angioendothelioma. Diagn Cytopathol, 2010; 39(7): 514–516.

Word count: 1883 Tables: 1 Figures: 4 References: 31

Access the article online: DOI: 10.5604/01.3001.0014.8653

Table of content: https://otorhinolaryngologypl.com/issue/13832

Competing interests: The authors declare that they have no competing interests.

Corresponding author: Sylwia Kropidłowska MD; Department of Otolaryngology and Maxillary Surgery, St. Vincent de Paul in Gdynia; Wójta Radtkego street 1, 81-334 Gdynia, Poland; E-mail: sylwiakropidlowska@int.pl

Cite this article as: Piotrowski S., Smialek U., Kropidlowska S.: Dąbska Tumor – a rare neoplasm of the paranasal sinuses. Case report and review of world literature; Pol Otorhino Rev 2021; 10 (2): 34-39