Pleomorphic adenoma of the nasal cavity – a case report

Gruczolak wielopostaciowy jamy nosowej – opis przypadku

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ABSTRACT:

Introduction: Pleomorphic adenoma is a benign tumor which occurs most commonly in the major salivary glands. It is very rare in the nasal cavity and may be misdiagnosed because of its uncharacteristic clinical and histopathological manifestation.

Case report: A case of a 42-year-old patient with pleomorphic adenoma on the lateral nasal wall has been presented. The tumor was accidentally found during FESS and septorhinoplasty. Initially, based on the histopathological examination of the obtained sample, there was a suspicion of adenoid cystic carcinoma. Later, the neoplasm was completely removed by the open septorhinoplasty and histopathological re-examination showed features of pleomorphic adenoma. Long term follow-up was necessary due to the potential risk of local recurrence.

KEYWORDS: benign tumors, nasal cavity, pleomorphic adenoma

STRESZCZENIE:

Wstęp: Gruczolak wielopostaciowy jest nowotworem łagodnym występującym najczęściej w śliniankach przyusznych. Bardzo rzadko pojawia się on w jamie nosa, co w połączeniu z jego niecharakterystycznym obrazem klinicznym i histopatologicznym stwarza ryzyko postawienia błędnego rozpoznania.

Opis przypadku: Przedstawiono przypadek 42-letniego pacjenta z gruczolakiem wielopostaciowym na bocznej ścianie nosa. Guz został wykryty przypadkowo podczas endoskopowej operacji zatok i septoplastyki, a na podstawie badania histopatologicznego pobranego wycinka początkowo wysunięto przypuszczenie raka gruczołowo-torbielowatego. Guz został doszczętnie usunięty z dostępu do septorynoplastyki otwartej. W ponownym badaniu histopatologicznym wykazano cechy gruczolaka wielopostaciowego. Ze względu na ryzyko wznowy miejscowej, pacjent pozostał pod opieką poradni laryngologicznej.

SŁOWA KLUCZOWE: gruczolak wielopostaciowy, jama nosowa, nowotwór łagodny

ABBREVIATIONS

FESS – functional endoscopic sinus surgery
MRI – magnetic resonance imaging

INTRODUCTION

Salivary gland tumors constitute a very diverse and heterogeneous group [1]. Their prevalence in the recent four decades has increased, however they are still very rare and constitute approx. 6% of diagnosed head and neck cancers [1, 2].

Pleomorphic adenoma, also referred to in the literature as mixed tumor, is the most frequent benign salivary gland tumor (it constitutes even up to 50% of all cancers of these organs) [3]. It particularly affects the salivary glands, and more rarely submandibular and sublingual glands. A total 35% of cases involve a localization in the minor salivary glands [4]. Both primary as well as the recurrent pleomorphic adenoma carries the risk of malignant transformation [5]. It usually occurs after 15–20 years in 5 to 25% of untreated patients [6] and can also potentially lead to the occurrence of distant metastases. The survival rates in such a situation are from 30 to 70% [5].

In addition to the typical localization, rare cases also involve pleomorphic adenoma developing within the hard and soft palate, lips, pharynx, larynx, trachea, in the lacrimal glands and in the external auditory canal [3, 4]. It is very rare for pleomorphic adenoma to occur in the nasal cavity, and whenever this happens, it most often occurs within the nasal mucosa [4].

Mixed tumor, both in its typical location as well as in the nasal cavity, is more prevalent in middle-aged women. The incidence

ratio of women to men is 3 to 2 [7, 8]. The most frequent symptoms accompanying pleomorphic adenoma of the nasal cavity include: nasal obstruction (58%) and occasional bleeding (26%) [4, 8].

Diagnostic challenges in mixed tumor of the nasal cavity are caused not only by the lack of characteristic clinical symptoms, but also nonspecificity in radiological imaging. Computed tomography usually reveals as a well-defined, homogeneous change. The presence of osteolysis may suggest a malignant nature of tumor. Magnetic resonance imaging may present a different picture, but it is usually clear. Signal intensity in the T1 sequence is low or moderate, and in the T2 sequence – high [9].

The final diagnosis is mainly based on the histopathological evaluation of the removed lesion [8]. The mixed tumor consists of epithelial and mesenchymal tissue. Compared to lesions in the salivary glands, tumors in the nasal cavity are characterized by a predominance of epithelial components and a lack of capsule. A significant predominance of epithelial cells in tumor tissue is possible, which may further hinder diagnosis due to similarity in the microscopic image to a malignant lesion. Histopathological diagnosis can be confirmed by immunohistochemical staining for the expression of cytokeratin, vimentin, and proteins: S100, SMA and GFAP, indicating the mixed nature of tumor. Excessive expression of p53, HER-2 and Ki-67 may indicate the presence of malignancy [9].

The core method of treating pleomorphic adenoma in the nasal cavities is complete surgical resection with a clean margin [8]. In most cases, it can be performed under endoscopic guidance [8, 10].

The risk of recurrence after surgical treatment of a tumor in the nasal cavity is relatively minor (8%), while it is more frequent when the tumor involves the paranasal sinuses [8]. Recurrences are locoregional and late, and usually exceed the two-year follow-up period [10].

CASE REPORT

A 42-yeard-old man was admitted to the Clinic of Otorhinolaryngology, Head and Neck Surgery of the Medical University of Warsaw in urgent mode, as part of a fast oncological therapy pathway, due to a right-sided nasal tumor.

The patient was diagnosed with the tumor accidentally during functional endoscopic sinus surgery (FESS) and septoplasty, performed 2 months earlier in a planned mode at another center for chronic sinusitis and deviated septum. FEES, which progressed uncomplicated, revealed an abnormal tissue mass about 20 mm in diameter located on the lateral wall of the nasal cavity near the inferior nasal concha; biopsies were taken for histopathological examination. Histopathological description, which was not unequivocal, involved the possibility of adenoid cystic carcinoma. Due to the raised suspicion of a malignant proliferative process in the nasal cavity, the patient was referred to the Otorhinolaryngology Clinic of the Medical University of Warsaw for qualification for further surgical treatment.

In physical examination performed at admission to the Clinic, the patient did not report any complaints related to the presence of a tumor. He negated headaches, recurrent nosebleeds, and leakage of pathological contents, reduced patency and impaired sense of smell. The patient was chronically treated for hypertension, which was well controlled and reduced with medication. The patient reported nicotinism estimated at 30 pack-years.

In ENT examination, the described change was not visible in anterior rhinoscopy. The nose was patent, and the mucosa was pink and completely healed after the procedure. There was a slight hypertrophy of the lower nasal conchas and a small amount of mucous discharge on the surface of the conchas. No leakage of pathological contents was found along the posterior wall of the pharynx.

As part of preoperative diagnostics, the patient underwent craniofacial MR with contrast, which described a solid-structured pathological change: upper-lower 20mm, two-sided 9.4 mm, anteroposterior 16 mm. The lesion was protruding and related to the anterior part of the lower right nasal concha with involvement of the head and perpendicular plate. Features of bone destruction of the marginal osseous lamina of the conche and thinning of the right nasal bone below the bridge of nose, directly at the lesion level, were also described. Infiltration outside the nasal cavity was not found.

After analyzing the results of imaging and histopathological examinations, the patient was qualified for surgical excision of tumor via open septorhinoplasty approach. Initially, the tumor was not visible in the nasal cavity, and the site of collected sample displayed the presence of adhesion of the nasal and septal mucosa. After incision of the junction of the quadrangular cartilage and the rim of piriform aperture, the nasal mucosa was widely dissected reaching the tumor. From the side of the nasal bone it had smooth outlines, while from the side of the mucosa the border was not clearly visible. During surgery, the lower nasal bone and lower anterior fragment of the frontal process of maxilla were excised with the tumor. Part of the nasal vault and nasal septum mucosa was also dissected. This was followed by removal of the anterior part of the inferior nasal turbinate and opening of the prelacrimal recess. The procedure was concluded with collection of marginal specimens from the mucosa of wound edges to assess the margins of the surgical incision.

The specimens taken during the surgical procedure were given for histopathological examination, which revealed a lack of morphological features typical of adenocarcinoma (no signs of neuroinvasion and destruction of cribriform structures). The whole morphological picture was described as closest to pleomorphic adenoma. The collected border specimens did not reveal neoplastic infiltration, confirming the radicality of surgical resection.

The patient was discharged from the hospital in good general condition on the 8th day after surgery.

In order to verify earlier suspicion of cystic-glandular cancer, the patient was recommended to provide previously made microscopic preparations and compare them with those currently performed. During histopathological consultation, the presence

of the previously described malignant tumor was excluded and the identical pattern of the lesions in the samples taken during endoscopy and in the preparation of the removed tumor was confirmed. The following features spoke of the picture of multiform adenoma: tumor-like pattern made of atypical cells, forming mainly cord-like and tubular patterns, and solid systems in places. Within the cord-like and tubular patterns, the luminous cells were larger, with a slightly follicular nucleus, immunohistochemically expressing strong expression of CK8/18; some of them showed the expression of CD117. Cells with myoepithelial morphology were located peripherally and presented strong p63 expression, S100 expression and weak CK8/18 expression. There were no current features of cell pleomorphism and visible subdivision figures (0/10 DPW), Ki67 proliferative activity was about 1%. There were visible foci of squamous cell differentiation with actinic keratosis and areas of mucoid, partially cartilaginous stroma. There was no tumor necrosis, or angioinvasive or neuroinvasive features in the material.

Due to the exclusion in histopathological examination of the excised malignant tumor, after an oncological consilium, the patient was referred to the ENT Clinic for further continuous observation.

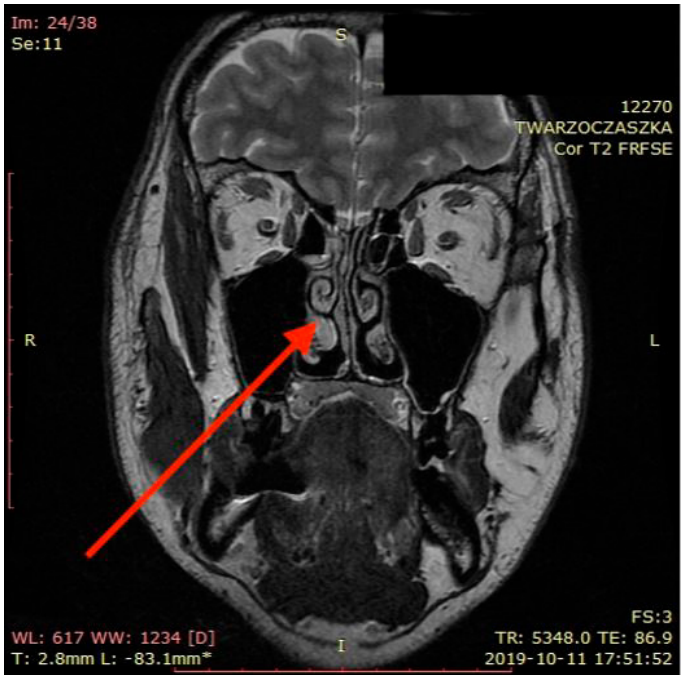

Fig. 1. Craniofacial MRI with contrast, which describes a tumor of dimensions: upper-lower 20 mm, two-sided 9.4 mm, anteroposterior 16 mm. The lesion was protruding and related to the anterior part of the lower right nasal concha with involvement of the head and perpendicular plate. The arrow indicates the tumor site.

DISCUSSION

Pleomorphic adenoma is extremely rare in the nasal cavity. If this is the case, it usually occurs within the nasal septum mucosa (reported frequency varies between 82.5% and 90%) despite the fact that mucous and serous glands are found mainly in nasal conchas located in the lateral wall of the nasal cavity [4]. The reason for such location remains unclear. A thorough literature review has shown that involvement by this tumor of the lower nasal conchas is very rare [11, 12]. In the presented case, pleomorphic adenoma was located on the lateral wall of the nasal cavity in the inferior nasal concha.

In the majority of reports, it was indicated that pleomorphic adenoma, both in its typical localization as well as in the nasal cavity, is more prevalent in women between the 3rd and 6th decade of life [7, 8, 11]. Most patients develop nonspecific symptoms such as nasal congestion, occasional nosebleeds, headache, and rhinitis [11]. The described patient did not report any of the above symptoms. Before the surgery he underwent craniofacial MRI with contrast, which described a solid lesion protruding and connected to the anterior part of the lower right nasal concha with involvement of the head and perpendicular plate. The largest superior-inferior one was 2 cm. This coincides with literature reports in which the size of the nasal multiform adenoma did not exceed 7.5 cm [13].

Preoperative diagnosis of pleomorphic adenoma in the nasal cavity, in contrast to that found in the salivary glands, is quite a challenge. Differential diagnosis should consider both benign and malignant tumors, such as squamous cell carcinoma, adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, inflammatory polyp, papilloma, hemangioma or osteoma [4, 14]. Diagnosis is to a large extent based on the result of histopathological examination, therefore proper collection of a specimen from the suspected lesion is key [8]. Preoperative histopathological examination also enables determination of whether a cancer has developed in the mixed tumor base in pleomorphic adenoma [5]. In the histological image, pleomorphic adenoma of the salivary glands is characterized by the presence of a thin collagen capsule and a clear demarcation between tumor and surrounding tissue. It consists of three components: tubuloductal, solid and mycosidal. Mixed tumor in the nasal cavity has an advantage of epithelial elements over connective tissue (stromal) and, unlike the mixed tumor in the parotid glands, it does not have a capsule. Epithelial cells are small, oval-shaped and often form cords.

A significant predominance of epithelial cells in tumor pattern is possible, with little or no stroma. This could additionally hinder diagnostics due to the similarity in the microscopic image to a malignant lesion, e.g. malignant mixed tumors [4, 9].

Diagnosis could be confirmed by immunohistochemical examination for the presence of expression of cytokeratin, vimentin, proteins: S100, SMA and GFAP, indicating the mixed nature of the tumor. Excessive expression of p53, HER-2 and Ki-67 may indicate the presence of malignancy [9].

The occurrence of infiltrating cancer cells and destructive growth patterns supports the diagnosis of pleomorphic adenoma [4, 9]. Differentiation of CxPA pleomorphic adenoma is challenging when all mixed tumor pattern undergoes malignant transformation or in the opposite situation – when the foci of tumor transformation (pleomorphic cells, frequent/atypical mitotic figures, hemorrhages, necrosis) are rare and disseminated [5, 15]. In such case, collecting a fragment of tissue located deeper within the tumor could increase the chances of a correct diagnosis [8]. The presented case involves a suspicion of glandular cystic cancer based on the histopathological

examination taken during FESS surgery. It is a slow-growing, malignant tumor originating from glandular tissue. The histological image of this tumor has tubular, cribriform and solid systems. Tumor cells are small, with sharp edges, scanty cytoplasm and with pleomorphic cell nuclei. They produce glandular-like structures based on matrix of glycosaminoglycans and basement membrane elements. Given the tumors found in the salivary glands, fine needle aspiration does not show adequate sensitivity in the diagnosis of this tumor, and accuracy is rated at 77%. Due to the similarity in the cytological image, it is sometimes confused with pleomorphic adenoma and less often with basal cell adenoma [16]. The final diagnosis of adenocarcinoma is possible on the basis of histopathological examination with immunohistochemical assessment – a positive reaction of tubular cells is observed in, among others CK5/6, CK7, CK8, CK18 and CD117. Myoepithelial cells react differently with calponin, S-100 protein, smooth muscle actin and p63 [16].

The core method of treating pleomorphic adenoma within the nasal cavity should be surgical treatment involving total tumor resection with a negative surgical margin [12]. Surgical access depends on the location and size of tumor and may include: lateral rhinotomy, eversion of facial layers, intranasal excision and partial-thickness mandibular resection [4, 8]. Some small-sized benign tumors of the nasal cavity can be successfully treated with endoscopic access [12]. In our case, the patient was qualified for surgery to excise the tumor via access used in open septorhinoplasty. The risk of recurrence after surgery described in the literature is relatively minor and amounts to 8% of cases [17]. Those are usually locoregional and late, exceeding the two-year observation period [10]. The risk of malignant transformation is 6%.

It is estimated to occur in 1.5% of patients who have not undergone resection within 5 years. Studies conducted by J. Compagno and R.T. Wong demonstrated that of 40 patients with pleomorphic adenoma within the nasal cavity who underwent surgery, only 7.5% (3 patients) had local recurrence after 3 years of observation [17, 18]. Constant follow-up of patients in ENT outpatient clinics is recommended.

CONCLUSIONS

Pleomorphic adenoma in the nasal cavity and particularly on the lateral wall, is extremely rare. It carries the risk of malignant transformation, therefore both benign and malignant tumors should be considered in differentiation. Diagnosis is based on the result of histopathological examination, which can be confirmed by immunohistochemical staining. The core method of treatment is total surgical resection. Long-term ENT follow-up is essential due to the risk of recurrence, which is usually locoregional and late.

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