Case report
Prikaz bolesnice

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Multifocal metachronous occurence of different hystologic sinonasal-type papilloma: a case report

Multifokalna metakrona pojava različitih histoloških sinonazalnih papiloma: prikaz bolesnice

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Summary

Introduction: Sinonasal-type papilloma is a very rare tumor, most commonly connected with the sinonasal space, and very rarely with the middle ear. Primary tumors of the temporal bone are extremely rare and only 28 cases have been described in literature, with additional 29 cases of tumor spreading from the sinonasal tract to the temporal bone.

Case presentation: We discuss the case of a 49-year-old woman who had a primary right-sided exophytic form of the sinonasal papilloma of the middle ear, which led to right-sided hearing loss, aural fullness, and otorrhea. During postoperative CT and MRI follow-up one year after surgery, a sinonasal oncocytic-type papilloma was discovered in the sphenoid sinus. To our knowledge, this is the first described case of histologically two different primary sinonasal-types of papilloma in a patient. Common presenting symptoms associated with sinonasal papilloma of the middle ear can be easily misdiagnosed with chronic otitis media or Eustachian tube dysfunction. Although primarily benign, sinonasal papillomas are locally aggressive and pose a risk of recurrence and malignant transformation. Therefore, surgery remains the treatment of choice with necessary long-term follow-up, to detect relapse or even a completely new tumor in that area.

Key words: exophytic type; middle ear; papilloma; sinonasal papilloma; sphenoid sinus; temporal bone

Sažetak

Uvod: Sinonazalni tip papiloma vrlo je rijedak tumor, najčešće vezan uz sinonazalni prostor, a vrlo rijetko za srednje uho. Primarni tumori temporalne kosti izuzetno su rijetki, a u literaturi je opisano samo 28 slučajeva, uz dodatnih 29 slučajeva širenja tumora iz sinonazalnog trakta u temporalnu kost.

Prikaz bolesnice: Prikazana je 49-godišnja žena koja je imala primarni desnostrani egzofitični oblik sinonazalnog papiloma srednjeg uha, koji je doveo do desnostranog gubitka sluha, osjećaja punoće u uhu i otoreje. Tijekom postoperativnog CT i MRI praćenja godinu dana nakon operacije, sinonazalni onkocitni tip papiloma otkriven je u sfenoidalnom sinusu. Prema našim saznanjima ovo je prvi opisani slučaj dvaju histološki različitih primarnih sinonazalnih tipova papiloma u jednog bolesnika. Uobičajeni simptomi povezani sa sinonazalnim papilomom srednjeg uha mogu se lako zamijeniti s kroničnom upalom srednjeg uha ili disfunkcijom Eustahijeve cijevi. Iako primarno benigni, sinonazalni papilomi su lokalno agresivni i predstavljaju opasnost od recidiva i maligne transformacije. Stoga operacija ostaje liječenje izbora, uz potrebno dugotrajno praćenje, kako bi se otkrio recidiv ili čak potpuno novi tumor na tom području.

Ključne riječi: egzofitični tip; srednje uho; papilom; sinonazalni papilom; sfenoidni sinus; temporalna kost

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Introduction

A rare tumor formed from the Schneiderian membrane, which lines the nasal cavity and paranasal sinuses, called sinonasal-type papilloma (formerly known as Schneiderian papilloma), most frequently develops in the sinonasal tract.[1] Although it has the same histological characteristics, it is extremely uncommon in the middle ear, mastoid[1–5], lacrimal sac, or nasopharynx.[6] Sinonasal papillomas are divided into three histological types: exophytic (ESP), oncocytic (OSP), and inverted (ISP)[3], which is the most common (60%).[6-10] While it is considered benign, it is locally aggressive and tends to invade adjacent structures. Disease incidence cannot be accurately calculated due to scarce reports in literature. Etiology remains unknown, with two hypotheses proposed: direct extension from the sinonasal cavity through the Eustachian tube or primary involvement of the middle ear due to the metaplastic changes in the middle ear mucosa.[4] Symptoms most commonly include otorrhea, aural fullness, hypoacusis, tinnitus, and otalgia.[5] It has been noted that sinonasal papillomas of the temporal bone have a higher recurrence and malignant transformation rate compared to papillomas in the sinonasal tract.[1,7] We describe a case of a 49-year-old female with a primary ESP located in the middle ear and mastoid with another primary OSP located in the sphenoid sinus, discovered on regular CT and MRI follow-up one year after the surgery.

Case presentation

A 49-year-old female presented to the ENT clinic with a complaint of aural fullness, otorrhoea, and pulsating tinnitus of the right ear. Patient history was significant for hearing difficulties and intermittent tinnitus lasting for 4 years. An otoscopic examination of the right ear showed a slightly extruded and hyperemic eardrum covered with a small polyp in posterior-inferior parts. Endoscopic rhinoscopy found no pathological changes in the mucosa. Pure tone audiometry showed conductive hearing loss with an air-bone gap of 55 to 75 db across middle and high frequencies on the right side. CT and MRI (Figures 1,2) of the temporal bones showed a process in the right cavum tympani with postcontrast imbibition extending towards the apex of the pyramid, foramen lacerum, and dura. There was no pathology in the nose and the paranasal sinuses. Due to the afore mentioned findings, a right-sided tympano-mastoidectomy was performed. Intraoperatively, the mastoid antrum was filled by tumorous tissue. The tumor filled the tympanic cavity and encapsulated the auditory ossicles.

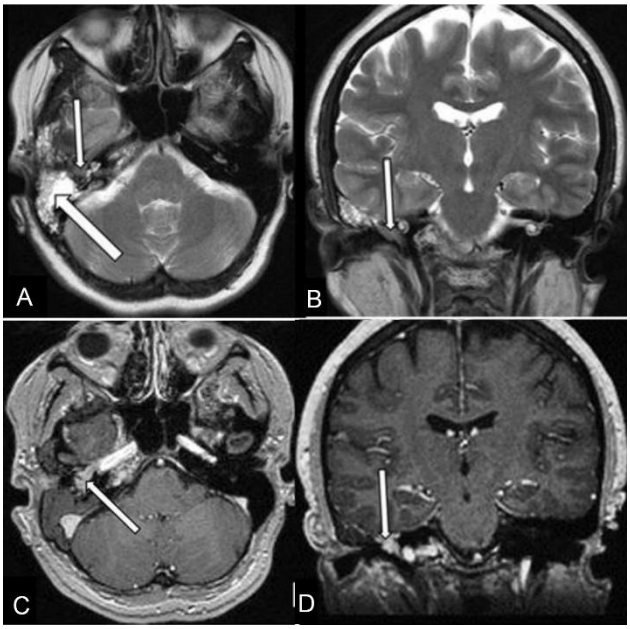

Figure 1 MRI of temporal bones, T2 images (A, B): A soft tissue formation in the right tympanum exhibiting a low signal (narrow arrow) and liquid content in the antrum and pneumatic cells of the right mastoid process exhibiting a high signal (wide arrow). In T1contrast enhanced images (C, D), a formation in the right tympanum can be seen, intensively imbibed with contrast (narrow arrow). The liquid content in the antrum and pneumatized mastoid cells does not show imbibition.
Slika 1. MRI sljepoočnih kostiju, T2 slike (A, B): formacija mekog tkiva u desnom bubnjištu s niskim signalom (uska strelica) i sadržaj tekućine u antrumu i pneumatskim stanicama desnog mastoidnog nastavka s visokim signalom (široka strelica). Na T1 prikazima s kontrastom (C, D) vidi se tvorba u desnom bubnjištu, intenzivno imbibirana kontrastom (uska strelica). Tekući sadržaj u antrumu i pneumatiziranim mastoidnim stanicama nije imbibiran.

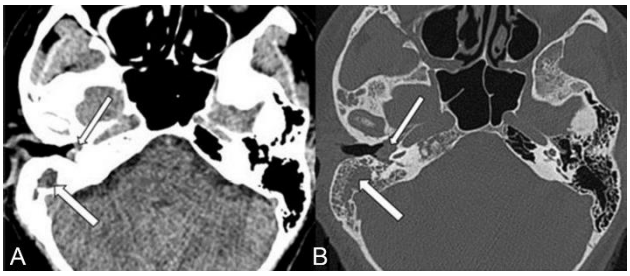

Figure 2 CT of the right temporal bone showing liquid content in the pneumatized right mastoid process and in the antrum. Imaging protocol for showing soft tissue structures (A) and imaging protocol for showing bone structures (B) was used. The right tympanum shows soft tissue content (narrow arrow).

Slika 2. CT desne sljepoočne kosti prikazuje tekući sadržaj u pneumatiziranom desnom mastoidnom nastavku i u antrumu. Korišten je protokol za prikaz struktura mekog tkiva (A) i slikovni protokol za prikaz koštanih struktura (B). Desno bubnjište prikazuje sadržaj mekog tkiva (uska strelica).

The incus and malleus were infiltrated by the tumor and were removed. The facial nerve canal and the Eustachian tube were not affected. Canal-wall-down tympanomastoidectomy was performed to facilitate complete disease removal from cavum tympani and epitympanum, safer follow-up and possible recurrence identification. Also, the tumor was removed from the foramen lacerum. The histopathology finding described an ESP. Immunohistochemical analysis was performed due to possible human papillomavirus (HPV) association and was HPV16 negative (Figure 3). One year after surgery, a control MRI and CT were performed and showed a new papilloma, located on the posterior part of the roof of the sphenoid sinus (Figure 4). Functional endoscopic sinus surgery (FESS) was performed and a small tumor was removed. The histopathology finding was the HPV16 positive OSP (Figure 3). Six months after the FESS, MRI showed no signs of recurrence. This case presentation has been assembled with informed consent from our patient, and IRB approval has been waived.

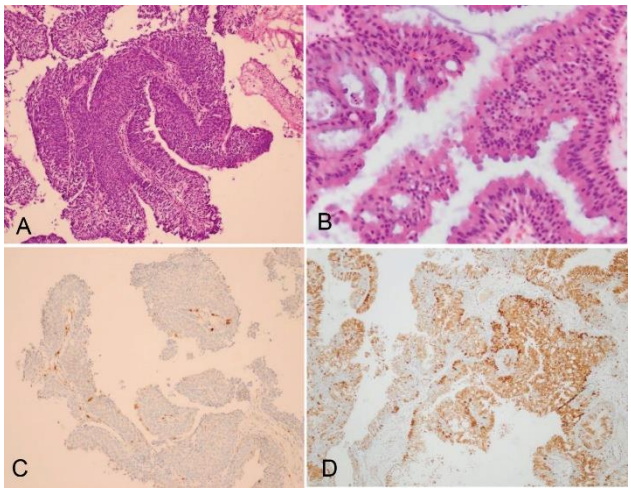

Figure 3 a) Exophytic papillary tissue consisting from villi, whose fibrovascular stroma was covered in reactive transitional, "Schneiderian" epithelium. According to WHO 2017 classification, it corresponds to an exophytic subtype papilloma (hematoxylin-eosin staining, 400× magnification). b) Exophytic papillary tissue consisting from villi, whose fibrovascularstroma was covered in oncocytic transitional, "Schneiderian" epithelium. According to WHO 2017 classification, it corresponds to an oncocytic subtype papilloma (hematoxylin-eosin staining, 400× magnification). c) Immuno-histochemical analysis was negative for possible human papilloma virus (HPV) association (p16 immunostaining, 200× magnification) d) Immunohistochemical analysis was positive on human papilloma virus (HPV) association (p16 immunostaining, 200× magnification).

Slika 3. a) Egzofitično papilarno tkivo koje se sastoji od resica, čija je fibrovaskularna stroma bila prekrivena reaktivnim prijelaznim, "Schneiderian" epitelom. Prema klasifikaciji WHO-a iz 2017. odgovara egzofitičnom podtipu papiloma (bojenje hematoksilin-eozinom, povećanje 400×). b) Egzofitično papilarno tkivo koje se sastoji od resica, čija je fibrovaskularna stroma prekrivena onkocitnim prijelaznim, "Schneiderian" epitelom. Prema klasifikaciji WHO-a iz 2017. odgovara onkocitnom podtipu papiloma (bojenje hematoksilin-eozinom, povećanje 400×). c) Imunohistokemijska analiza bila je negativna na moguću povezanost humanog papiloma virusa (HPV) (p16 imunološko bojenje, povećanje 200×) d) Imunohistokemijska analiza bila je pozitivna na povezanost humanog papiloma virusa (HPV) (p16 imunobojenje, povećanje 200×).

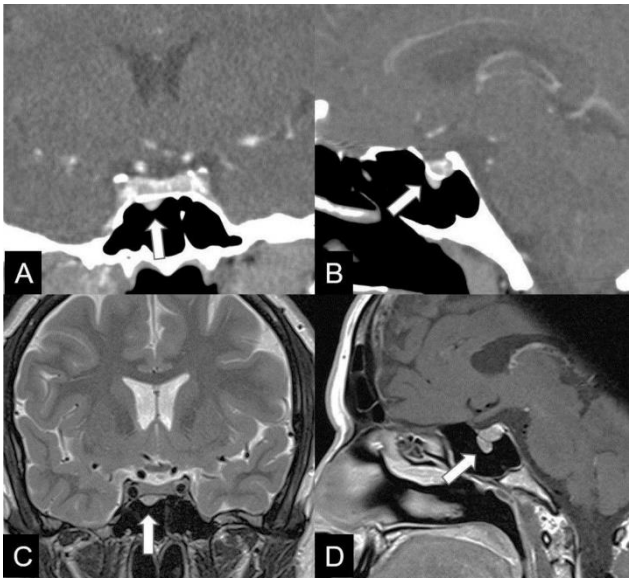

Figure 4 Coronal and sagittal CT images (A, B) demonstrate a well-defined solid mass (arrows) with homogeneous enhancement in the sphenoid sinus roof without defect of bony contour of sphenoid sinus. Coronal T2WI (C) and sagittal contrast-enhanced T1WI (D) MRI images show marked enhancement of the tumor in the sphenoid sinus roof (arrows).

Slika 4. Koronalne i sagitalne CT slike (A, B) pokazuju dobro definiranu čvrstu masu (strelice) s homogenim pojačanjem u krovu sfenoidnog sinusa, bez defekta koštane konture sfenoidnog sinusa. Koronalne T2WI (C) i sagitalne T1WI (D) MRI slike pokazuju značajno povećanje tumora u krovu sfenoidnog sinusa (strelice).

Discussion

So far, only 29 cases of the primary sinonasal papilloma affecting the temporal bone have been

reported. Recurrence, according to the majority of authors, indicates an insufficient resection and frequently occurs at the initial site within two years following surgery. The multifocal appearance of the same type of sinonasal papilloma has been described.[6] To our knowledge, this is the first described case of different types of sinonasal-type papilloma (exophytic and oncocytic) in two different locations (middle ear and sphenoid sinus) in a patient. The ESP in the middle ear, which was identified in our case is very rare and only four cases described to date. In our case, direct extension from the sinonasal tract can be excluded since the tumor was localized in the mastoid and middle ear space, while an extension to the Eustachian tube was not present. Clinical presentation mostly resembles chronic otitis media with polyp formation in the posterior tympanic membrane segments, but a glomus tympanicum should also be considered, especially if there is brisk bleeding present upon manipulation[6], as well as Eustachian tube dysfunction.[7,8] The etiology is probably multifactorial – no cases so far have been unequivocally linked with HPV infection, including ours, yet its contribution cannot be ruled out completely.[6] Histological features are similar to sinonasal localized Schneiderian-type papillomas. The characteristic features of ESP - fibrovascular stroma covered in the reactive transitional epithelium were found in our case. Microcysts, muciphages, goblets cells, and an inflammatory infiltration may also be present in Schneiderian-type papillomas.[9] A recent report showed that ESP does not show malignant transformation, but its biological behavior in the middle ear is difficult to predict.[3] Recurrence was reported at 100% in all previously reported cases treated only with tympanoplasty and simple excision compared to 39% following radical surgery (mastoidectomy or temporal bone resection).[7,8] These results can be compared with recurrence rates of ISP when treated with a similar extent of surgery.[1] The treatment of choice is surgery–radical tympanomastoidectomy, with other approaches proving inefficient in disease control.[1] If the diagnosis of the tumor is done very early, like the second tumor in this case, then more preservative surgery can be performed. The tumor in the sphenoid sinus was still asymptomatic and was diagnosed only due to regular MRI follow-ups. Since it was a small tumor, it was easy to remove it using FESS. Sinonasal-type papillomas are very prone to recurrence but the occurrence of different histological types is not common. The diagnosis is usually obtained with CT, MRI, and radical surgery, due to the advanced stage of the tumor being the treatment of choice. Strict postoperative follow-up is necessary and should include routine otoscopy, nasendoscopy, and imaging due to the high possibility of recurrence, unknown malignant potential, and the lack of accurate prognostic indicators.

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