C A S E R E P O R T S

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Middle turbinate angiofibroma in an elderly woman

Angiofibrom srednje nosne školjke kod starije žene

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Abstract

Background. Angiofibromas are histologically benign vascular tumors, originating from the nasopharynx, near by the area of sphenopalatine foramen. These neoplasms occur typically in male adolescents. Reports of primary extranasopharyngeal angiofibromas have appeared sporadically in the literature in English. We present the first case of an elderly woman with tumor arising from the middle turbinate, diagnosed as angiofibroma. **Case report.** A 63-year-old female presented with left-sided nasal obstruction and epistaxis. Endoscopic evaluation revealed a polypoid mass arising from the anteroinferior portion of the left middle turbinate. Computed tomography (CT) scan showed a soft-tissue opacity that filled the anterior part of the left nasal cavity. After the endoscopic excision of the mass, postoperative pathohistological and immunohistochemical analysis confirmed the diagnosis of an angiofibroma. Two years later, the patient was free of symptoms and without endoscopic evidence of recurrence. **Conclusion.** Extranasopharyngeal angiofibromas arising from the nasal cavity are extremely rare tumors. Immunohistochemical analysis is very important in all doubtful cases, especially in those with atypical location.

Key words:
arteriovaenous malformations; turbinates; otorhinolaryngologic surgical procedures; immunohistochemistry.

Apstrakt

Uvod. Angiofibromi su histološki benigni, vaskularni tumori, poreklom iz nazofarinksa, neposredno iz područja sfenopalatinskog gangliona. Tipično se javljaju kod adolescenata muškog pola. Slučajeve primarno ekstranazofaringealnih angiofibroma retko nalazimo u literaturi. Mi predstavljamo prvi registrovani slučaj starije žene sa tumorom koji je polazio iz srednje nosne školjke, dijagnostikovanim kao angiofibrom. **Prikaz bolesnika.** Žena, stara 63 godine, javila se sa otežanim disanjem na nos i krvarenjima iz leve strane nosa. Endoskopski pregled pokazao je prisustvo polipoidne mase koja je polazila sa prednje-donjeg dela leve srednje nosne školjke. Kompjuterizovana tomografija (KT) pokazala je mekotkivnu masu koja je ispunjavala prednji deo leve šupljine nosa. Nakon endoskopske ekscizije tumora, postoperativne patohistološke i imunohistohemijske analize potvrdile su dijagnozu angiofibroma. Dve godine kasnije, bolesnica je bila bez simptoma i bez recidiva tumora. **Zaključak.** Ekstranazofaringealni angiofibromi, primarno organizovani u nosnoj duplji ekstremno su retki tumori. Imunohistohemijska analiza je veoma bitna u svim nejasnim slučajevima, naročito u onim sa atipičnom lokalizacijom.

Ključne reči:
arteriovenske malformacije; nos, konhe; hirurgija, otorinolaringološka, procedure; imunohistohemija.

Introduction

Angiofibromas are highly vascular, nonencapsulated, histologically benign but locally aggressive tumors which most commonly arise in the nasopharynx of adolescent males [1]. It is a unique fibrovascular tumor with the specific histopathological finding of irregulary configured endothelial lined vascular spaces embedded in a fibrous stroma [2]. They usually arise from the posterolateral wall of the nasal cavity, where the sphenoidal process of the palatine bone meets the horizontal ala of the vomer and the pterygoid process [1, 3, 4]. Angiofibromas constitute about 0.5% of all head and neck neoplasms [1, 3]. These tumors may rarely localise in extranasopharyngeal sites. To our knowledge, we present the first reported case of middle turbinate angiofibroma in an elderly female.

Case report

A 63-year-old woman came to the outpatient department with an eight month history of progressive left sided nasal obstruction and intermittent epistaxis. Five days be-

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fore our examination, the patient had two episodes of intensive nasal bleeding. Except the arterial hypertension which was medically controled, the patient had no other health problems. She referred to regular menstrual cycles, two normal pregnancies, and during the beginning of the symptoms, due to her age, she was in menopause for a long time. She had never use hormonal replacement therapy. The coagulation status was normal. Anterior rhinoscopy and endoscopic evaluation revealed a fragile, lobular, red-grayish colored, smooth, polypoidal mass, arising from the anteroinferior portion of the left middle turbinate, which bottom tip was above the lower end of the inferior turbinate. Posterior rhinoscopy was normal in appearance. Computed tomography (CT) scan of the nose and paranasal sinuses demonstrated a soft tissue opacity that filled the anterior part of the left nasal cavity, extending from the septum to the lateral nasal wall, without any sinus invasion and bony destruction (Figure 1). We decided to undertake

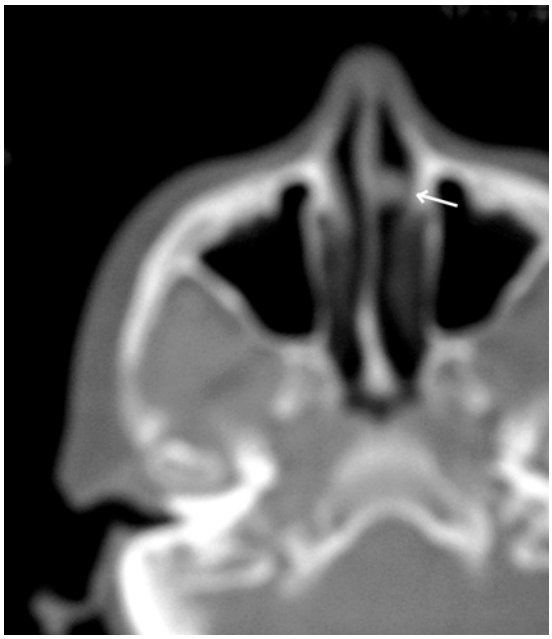

Fig. 1 – Axial computerized tomography (CT) scan showing a soft-tissue mass in the anterior part of the left nasal cavity extending from the septum to the lateral nasal wall

biopsy in epimucous local anaestesia. In attempt to biopsy tumor, we excided almost complete mass. It was lobular, smooth, red-grayish, about 15 mm long tumour, with the diameter up to 9 mm. This was followed by profuse bleeding which was controlled with anterior intranasal packing. The patient then underwent endoscopic endonasal examination in the operation theatre, under general anaesthesia. We found a small lobular lesion on the anterior part of the left middle turbinate. It had a thin pedicle which was bleeding intensively. After we resected the anterior third of the middle turbinate together with tumor, bleeding significantly decreased. The blood loss was estimated at 300 mL. The antibiotic therapy was used for ten days. The anterior nasal pack was removed on the fifth postoperative day and endoscopic nasal finding was good.

Histopathological examination of the excised masses (Hematoxylin-Eosin stain) showed a metaplastic squamous epithelium with the respiratory epithelium remnants on the tumor surface (Figure 2). Under the epithelium, we found many irregular blood vessels ranging from capillaries and sinusoids to large vessels, often with stellate or “staghorn” appearance, lined with one layer of flat endothelial cells lying in a fibrous stroma (Figure 2). Several fields of proliferate fibrosis at the sites of repetitive haemorrhages were seen (Figure 3). This suggested the diagnosis of angiofibroma.

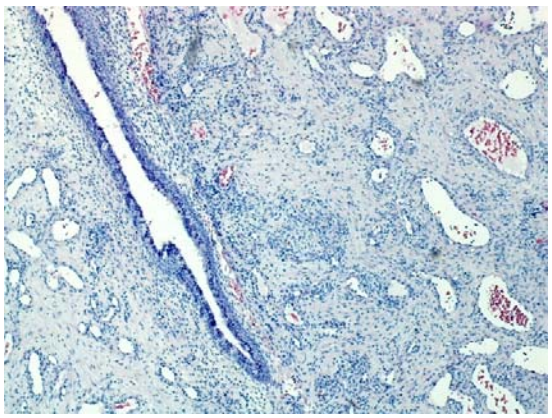

Fig. 2 – Angiofibroma: numerous blood vessels raning form capillaries and sinusoids to large vessels, lying in a fibrous stroma. Respiratory epithelium remnant in the tumor subsidence (H&E; 40x)

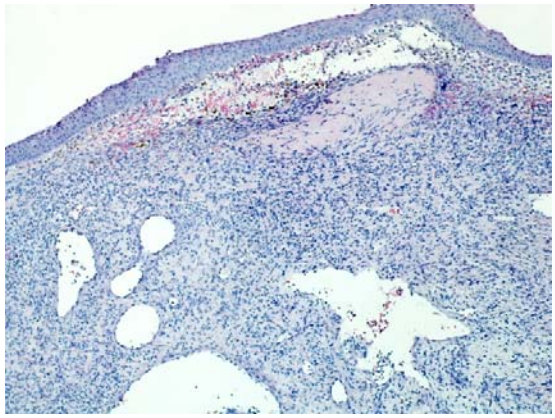

Fig. 3 – Angiofibroma: metaplastic squamous epithelium was present on the tumor surface; in the stroma, a field of proliferate fibrosis at the site of previous repetitive hemorrhage was seen (H&E; 40x)

Immunohistochemical analysis confirmed the diagnosis. We found a negative reaction for smooth muscle actin and desmin, and a positive reaction for stromal cells vimentin. All vascular cells were positive for CD34.

A CT scan with contrast, performed two months after the surgical treatment, discovered no residual tumor. Two years later, the patient was free of symptoms and without endoscopic and CT evidence of recurrence.

Discussion

Histologically, angiofibromas of the head and neck are benign non-encapsulated lesions occuring predominantly in the nasopharynx [3]. From its origin in the sphenopalatine foramen region, the tumor grows in all directions through multiple projection [4]. Another controversial characteristic is its tendency to grow through high resistance ways between the bones, instead of only occupying space [4]. Some authors believe that the tumor aggressivness is related to the patient's age, observing that both intracranial and infratemporal invasion usually occur in younger patients [4].

A number of theories have been proposed to explain the etiopathogenesis of angiofibroma, such as developmental, hormonal and genetic, but none of them found general acceptance. The current hypothesis, which categorizes juvenile angiofibroma as a vascular malformation, arose from the observation of discontinuous vascular basal laminae, focal lack of pericytes and pronounced irregularity of the smooth muscle layers [2]. In accordance with these morphological findings, the embriological explanation for the formation of this vascular tumor component is an incomplete regression of the first branchial arch artery [2, 5]. The predilection for adolescent males suggests a relationship with sex hormones [3]. Hormonal and growth factor receptors have been observed in the tumor tissue [6, 7]. A hormonal theory suggests that high estrogen levels protect females [7]. This could explain the male predominance and the small number of reported female cases. It has been hypothesized that angiofibroma is a testosterone-dependent tumor that arises from a fibrovascular nidus in the nasopharynx that lies dormant until the onset of puberty [7]. At this time, the tumor grows and becomes symptomatic. In the present case, the occurrence of angiofibroma after menopause may also support the idea of this hormonal influence. We supposed that our patient's tumor went through natural regression during her years of increased estrogen production and then started proliferation after menopause with its decrease hormonal effect. However, a case of angiofibroma in a pregnant woman reported by Peloquin et al. [8] favors the possibility of other contributing factors in the etiopathogenesis of tumor.

According to Brunner [9], these tumors originate in the tissue of the anterior margin of the atlas at the lower surface of the sphenoid bone, which is called fascia basalis. During its developpment, this tissue extends up to the posterior part of the vomer and the ethmoid bone [9]. We suggested that our patient's tumor had arisen from the periosteum of the middle turbinate which is the part of ethmoid bone. We supposed that the fascia basalis existed in the remnants of the middle turbinate region. The presence of this tumor on the anterior portion of the concha media indicated that the origin may be from ectopic tissues located furter away to its usual place.

Reports of primary extranasopharyngeal angiofibromas have appeared sporadically in the literature. They most commonly originate from the maxillary sinus [1, 3]. The ethmoid and sphenoid sinuses, nasal septum, middle and inferior turbinates, conjunctiva, molar and retromolar region, tonsil and larynx are other sites where extranasopharyngeal angiofibromas have been reported [1, 3, 10]. Recently a group of 65 patients with extranasopharyngeal angiofibromas has been reported by Windfuhr and Remmert [3]. The oldest patient was 78 years old. The mean age of presentation in these extranasopharyngeal angiofibromas was 23 years, in comparison to the mean age of presentation in nasopharyngeal angiofibromas which was 17 years [3]. Unlike nasopharyngeal angiofibromas, extranasopharyngeal angiofibromas occur more frequently in females at later ages [3].

In Medline® and Google™ search, we could find only two cases of middle turbinate angiofibroma. The first case was reported by Peloquin et al. [8] in a 31-year-old pregnant woman. The tumor had a pedicle on the inferoposterior portion of the right middle turbinate, near the sphenopalatine foramen [8]. The second one was reported by Huang et al. [11] in a 14-year-old boy. The mass was seen to arise from the medial surface of the right middle turbinate, near its attachment to the cribriform plate [11]. In our patient case, the tumor pedicle was on the anterior portion of the middle turbinate. However, we could not find any case of the middle turbinate angiofibroma in an elderly woman in the literature in English.

The most frequent symptoms of nasopharyngeal angiofibromas are unilateral nasal obstruction, epistaxis, facial deformities and pain. Extranasopharyngeal angiofibromas symptoms are related to their site of origin [1, 3].

Optimal management of angiofibromas includes preoperative radiological examination and biopsy [3, 10]. Computerized tomography MRI and arteriography are valuable diagnostic procedures in the evaluation of nasopharyngeal angiofibromas [3, 12]. Selective arteriography clearly demonstrates vascular pattern and blood flow dinamics and allows preoperative selective embolisation to reduce intraoperative bleeding [3, 12]. However, the exclusion of hypervascularity with arteriography does not exclude extranasopharyngeal angiofibromas [3]. Computerized tomography, is sufficient for diagnosis, because it clearly delineates and identifies the tumor [3]. Endoscopic examination is warranted, but biopsy is ill-advised without adequate plans for hemostasis [3, 10].

Various modalities have been used for treatment of angiofibromas, including surgery, hormonal therapy, radiation and systemic chemotherapy [3, 10]. However, surgery remains the primary course of treatment [3, 5]. Surgical procedures include transpalatal techniques, lateral rhinotomy, midfacial degloving, infratemporal aproaches and combined infratemporal and frontotemporal techniques [5]. The evolution of endoscopes, special instruments and experience in sinus surgery allows for minimal invasive endonasal resection of angiofibromas today [5]. Advantages of the endoscopic technique are minimal invasive character and low morbidity [5, 13]. In our case, location and dimension of tumor permitted us to remove it completely, using a simple endoscopic surgical procedure, without previous selective arteriography and embolisation.

Atipical location of angiofibroma needs a precise differential diagnosis in relation to the fibrosed antrochoanal and ethmoidal polyp and other fibrovascular tumors, such as capillary hemangioma, hemangiopericytoma and solitary fibrous tumor [14, 15]. A positive reaction to vimentin and CD34, and negative reaction to actin and desmin found at immunohistochemistry is sufficient for diagnosis of extranasopharyngeal angiofibroma.

Conclusion

In conclusion, a cause of angiofibroma remains unknown. Extranasopharyngeal angiofibroma arising from the nasal cavity is an extremely rare tomor. It probably comes from an ectopic tissue. Surgical excision of the mass is the treatment of choice. Immunohistochemical analysis is very important in all doubtful cases, especially in those with atypical location.

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