Case Report / Olgu Sunumu

Extranasopharyngeal angiofibroma of the posterior nasal septum: a rare clinical entity

Posterior nazal septumun ekstranazofarengeal anjiyofibromu: Nadir bir klinik oluşum

Sinan Atmaca, M.D.,[1] Cem Bayraktar, M.D.,[1] Levent Yıldız, M.D.[2]

[1]*Department of Otolaryngology, Medicine Faculty of Ondokuz Mayıs University, Samsun, Turkey*
[2]*Department of Pathology, Medicine Faculty of Ondokuz Mayıs University, Samsun, Turkey*

Angiofibroma of extranasopharyngeal origin is very rare. Although it is usually originated from any mucosal structure in the head and neck region, maxilla is the most common involvement site. The nasal septum is an exceptional anatomic site of an angiofibroma. Surgery is the best treatment modality and recurrence is very rare. Nasal septal angiofibromas must be considered in the differential diagnosis of nasal vascular masses arising from the nasal septum. In this article, we report a 37-year-old male case with nasal septal angiofibroma who underwent surgical resection of the tumor. This is the 16th case in the literature.

Key Words: Angiofibroma; extranasopharyngeal angiofibroma; nasal septum.

Ekstranazofarengeal yerleşimli anjiyofibrom çok nadir görülmektedir. Genellikle baş ve boyun bölgesindeki herhangi bir mukozal yapıdan köken almasına rağmen, maksilla en sık görülen tutulum bölgesidir. Nazal septum, anjiyofibromun istisnai bir anatomik bölgesidir. Cerrahi en iyi tedavi seçeneği olup, nüks çok azdır. Nazal septal anjiyofibromlar, nazal septumdan köken alan vasküler kitlelerin ayırıcı tanısında göz önünde bulundurulmalıdır. Bu yazıda, nazal septal anjiyofibromu olan ve tümörü cerrahi olarak rezeke edilen 37 yaşında bir erkek olgu sunuldu. Bu, literatürdeki 16. olgudur.

Anahtar Sözcükler: Anjiyofibrom; ekstranazofarengeal anjiyofibrom; nazal septum.

Juvenile nasopharyngeal angiofibromas (JNAs) are seen in the lateral wall of nasopharynx and patients are males usually aged between 7-25 years.[1,2] Rarely, angiofibromas can occur outside the nasopharynx. These groups of angiofibromas are called extranasopharyngeal angiofibromas (ENAs). Patients with ENAs are commonly females and older patients compared to those with JNAs.[2-5] In the head and neck region, they can originate from any mucosal structure.[1] The most common site in many studies is the maxilla.[2,4-8] Less frequently, they can also arise from the nasal septum, turbinates, ethmoid cells, tonsils, cheek, larynx, sphenoid, retromolar area

Available online at
www.kbbihtisas.org
doi: 10.5606/kbbihtisas.2013.29795
QR (Quick Response) Code

Received / *Geliş tarihi:* October 31, 2012 Accepted / *Kabul tarihi:* December 12, 2012

Correspondence / İletişim adresi: Sinan Atmaca, M.D. Ondokuz Mayıs Üniversitesi Tıp Fakültesi Kulak Burun Boğaz Hastalıkları Anabilim Dalı, 55139 Kurupelit, Samsun, Turkey. Tel: +90 362 - 312 19 19 e-mail *(e-posta):* sinanatmaca@yahoo.com

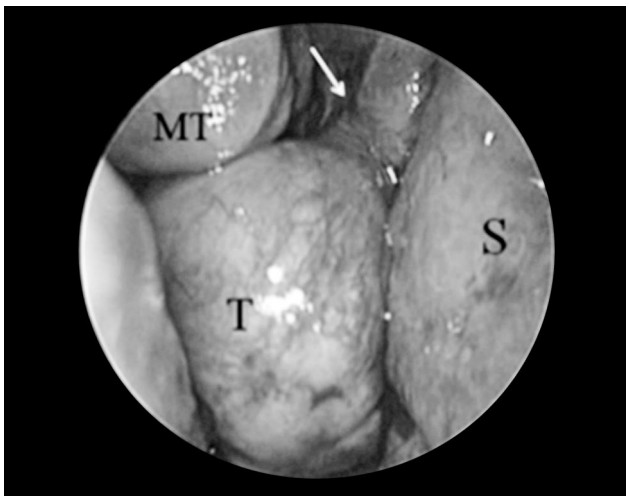

Figure 1. *Purple-reddish nasal mass filling the right nasal choana. MT: Middle turbinate; S: Nasal septum; T: Tumor; Arrow: Stalk of the tumor.*

and infratemporal fossa.[5,9-12] We report what is to our knowledge the 16th case of nasal septal angiofibroma in the literature.

CASE REPORT

A 37-year-old male presented to our outpatient clinic with a history of long-term right-sided nasal obstruction that progressed within the last six months and epistaxis for the last three-four years. Epistaxis was occasional and he denied any history of bleeding disorder. Endoscopic examination revealed a purple-reddish mass filling the nasopharynx that was anchored to the posterior nasal septum with a stalk (Figure 1). Contrast enhanced computed tomography showed a mass arising from the vomerian septum lacking prominent enhancement with regular borders, and no expansion to the paranasal sinuses (Figure 2).

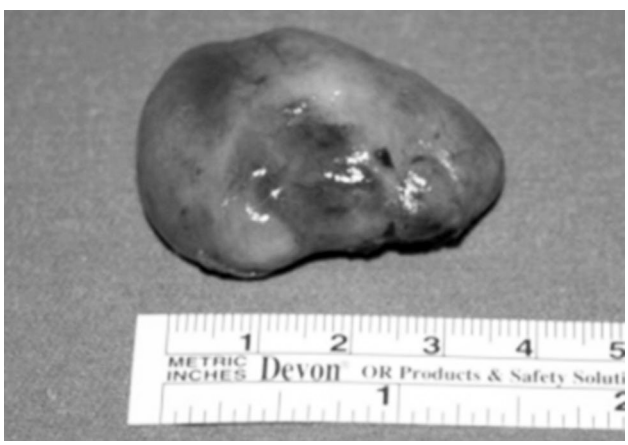

Figure 3. *Excisional biopsy specimen of the nasal mass.*

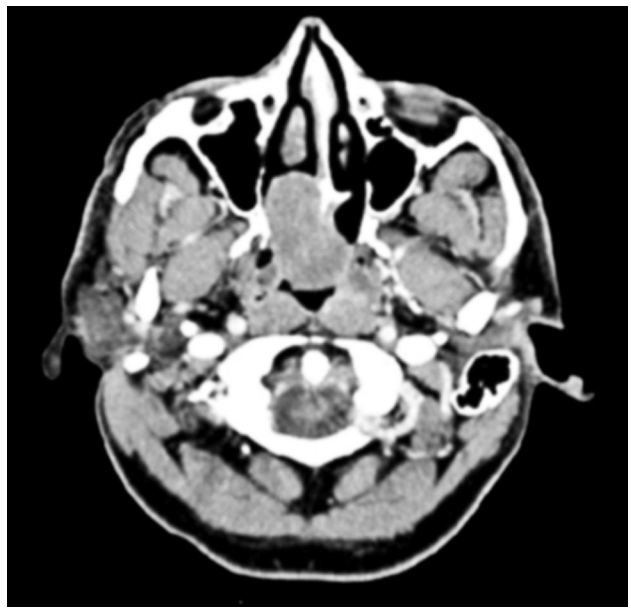

Figure 2. *Axial computed tomography scanning image of the posterior nasal septal mass.*

The patient was taken to the operating room for surgical management. Under general anesthesia, the posterior nasal septum and the stalk were infiltrated with 2% lidocaine hydrochloride. The mass with underlying mucoperiosteum was elevated off the septum with a 5 mm safety margin and the stalk was excised with bipolar cautery at the base of the lesion. Blood loss was minimal. A biopsy specimen of the mass measuring 3x4 cm was obtained (Figure 3). Anterior nasal packing was done and removed after 48 hours. The patient's postoperative recovery was uneventful. Histopathological examination revealed an angiofibroma. Hematoxylin and eosin

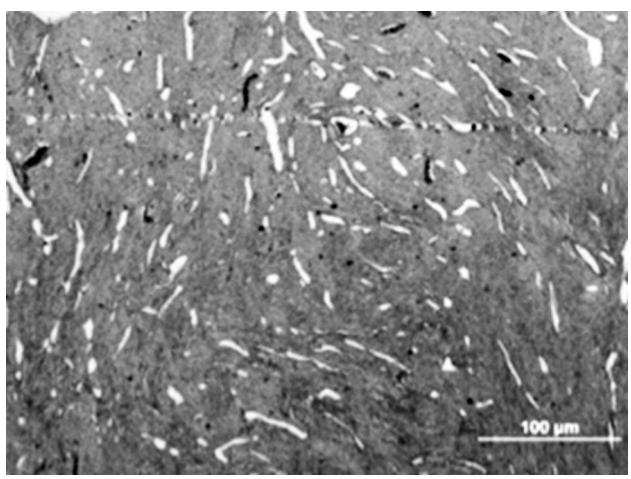

Figure 4. *Multiple dilated, thin-walled vascular spaces separated by dense stroma (H-E x 25).*

stains displayed multiple thin-walled and dilated vascular spaces separated by fibroblastic, cellular and dense stroma containing excessive collagen (Figure 4). Follow-up at seven months revealed no sign of recurrence.

DISCUSSION

Extranasopharyngeal angiofibromas must be examined as a separate clinical entity from JNAs.[5,8] Nasal septal angiofibromas (NSAs), which can be defined as a subgroup of ENAs, are very rare and only 15 cases have been reported until now. The most common presenting symptoms are epistaxis and nasal obstruction. At first glance; JNAs and NSAs may look similar but when compared to each other, patients with NSA are older, sex predilection of male gender with JNA doesn't apply to NSA in which females can also be affected and hypervascularity is less prominent with NSAs.[8,13] Computed tomography (CT) and MRI can be helpful in the diagnosis of NSAs both by determining the tumor size and its extension and providing information about vascularity of the tumor. Nasal septal angiofibromas show mild or no enhancement on CT and MRI due to poor vascularity as opposed to strong enhancement by hypervascular JNAs.[8,13] Our patient also showed no significant enhancement on the CT scan. Nasal septal angiofibroma was first described by Hiraide and Matsubara[7] in 1984. They reported on a 13-year-old male with a history of recurrent epistaxis and left-sided nasal obstruction having a nasal mass attached to the anterior two third of the nasal septum with a small pedicle. After Hiraide and Matsubara,[7] several authors reported nasal septal angiofibromas.[3,6,8,13-18] Two theories have been proposed to explain the pathogenesis of NSAs. The first theory suggests that the tumor arises from the junction of the bony and cartilaginous septum where the origin of the angiofibroma is the periosteum of the perpendicular plate of the ethmoid bone which is caused by an abnormal anterior extension of the fascia basalis located in the posterior part of the vomer and the ethmoid bone.[5-7,19] However, the anterior NSAs can't be explained with this theory. The second theory proposes the existence of a turbinate-like vascular tissue that is retained as an ectopic nidus in developing periosteum during nasal septal growth.[6,20] Biopsy is not recommended for septal angiofibromas as with JNAs due to the risk of brisk bleeding. Somdaş et al.[17] reported better results with preoperative embolization for the excision of the septal angiofibroma via the open technique. Surgery is the best treatment modality and recurrence is very rare.[4,13] Nasal septal angiofibromas must be considered in the differential diagnosis of nasal vascular masses arising from the nasal septum.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Celik B, Erisen L, Saraydaroglu O, Coskun H. Atypical angiofibromas: a report of four cases. Int J Pediatr Otorhinolaryngol 2005;69:415-21.
2. Huang RY, Damrose EJ, Blackwell KE, Cohen AN, Calcaterra TC. Extranasopharyngeal angiofibroma. Int J Pediatr Otorhinolaryngol 2000;56:59-64.
3. Akbas Y, Anadolu Y. Extranasopharyngeal angiofibroma of the head and neck in women. Am J Otolaryngol 2003;24:413-6.
4. Castillo MP, Timmons CF, McClay JE. Autoamputation of an extranasopharyngeal angiofibroma of the nasal septum. Int J Pediatr Otorhinolaryngol Extra. 2006;1:267-70.
5. Windfuhr JP, Remmert S. Extranasopharyngeal angiofibroma: etiology, incidence and management. Acta Otolaryngol 2004;124:880-9.
6. Handa KK, Kumar A, Singh MK, Chhabra AH. Extranasopharyngeal angiofibroma arising from the nasal septum. Int J Pediatr Otorhinolaryngol 2001;58:163-6.
7. Hiraide F, Matsubara H. Juvenile nasal angiofibroma: a case report. Arch Otorhinolaryngol 1984;239:235-41.
8. Tasca I, Compadretti GC. Extranasopharyngeal angiofibroma of nasal septum. A controversial entity. Acta Otorhinolaryngol Ital 2008;28:312-4.
9. Dere H, Ozcan KM, Ergul G, Bahar S, Ozcan I, Kulacoglu S. Extranasopharyngeal angiofibroma of the cheek. J Laryngol Otol 2006;120:141-4.
10. Lerra S, Nazir T, Khan N, Qadri MS, Dar NH. A case of extranasopharyngeal angiofibroma of the ethmoid sinus: a distinct clinical entity at an unusual site. Ear Nose Throat J 2012;91:E15-7.
11. Nomura K, Shimomura A, Awataguchi T, Murakami K, Kobayashi T. A case of angiofibroma originating from the inferior nasal turbinate. Auris Nasus Larynx 2006;33:191-3.
12. Szymańska A, Szymański M, Skomra D, Szczerbo-Trojanowska M. Extranasopharyngeal angiofibroma of the infratemporal fossa. Otolaryngol Head Neck Surg 2009;140:433-434.
13. Garcia-Rodriguez L, Rudman K, Cogbill CH, Loehrl T, Poetker DM. Nasal septal angiofibroma, a subclass of

extranasopharyngeal angiofibroma. Am J Otolaryngol 2012;33:473-6.
14. Mohamed AA. Angiofibroma of the nasal fossae: apropos of 12 cases observed in Mali. Med Trop (Mars) 1994;54:247-8. [Abstract]
15. Mohindra S, Grover G, Bal AK. Extranasopharyngeal angiofibroma of the nasal septum: a case report. Ear Nose Throat J 2009;88:E17-9.
16. Sarpa JR, Novelly NJ. Extranasopharyngeal angiofibroma. Otolaryngol Head Neck Surg 1989;101:693-7.
17. Somdas MA, Ketenci I, Unlu Y, Canoz O, Guney E. Extranasopharyngeal angiofibroma originating from the nasal septum. Otolaryngol Head Neck Surg 2005;133:647.
18. Uyar M, Turanli M, Pak I, Bakir S, Osma U. Extranasopharyngeal angiofibroma originating from the nasal septum: a case report. Kulak Burun Bogaz Ihtis Derg 2009;19:41-4.
19. Brunner H. Nasopharyngeal fibroma. Ann Otol Rhinol Laryngol 1942;51:30-65.
20. Schiff M. Juvenile nasopharyngeal angiofibroma. a theory of pathogenesis. Laryngoscope 1959;69:981-1016.