



Extranasopharyngeal angiofibroma originating from the nasal septum: a case report

Nazal septumdan kaynaklanan ekstranasofarengal anjiyofibrom: Olgu sunumu

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Angiofibromas (AFs) originate predominantly in the nasopharynx and posterior nasal cavity. The incidence of these lesions is less than 0.5% among all neoplasms of head and neck. They originate outside the nasopharynx rarely and can therefore be misdiagnosed. The nasal septum is an extremely rare site. Only four cases of extranasopharyngeal angiofibromas arising in the nasal septum have been reported so far. We report here a case of a vascular mass arising from the nasal septum in a 19-year-old boy which was extirpated by submucosal resection endonasally. Histopathology confirmed it to be a case of angiofibroma.

Key Words: Angiofibroma; nasal septum; nasopharyngeal.

Anjiyofibromlar (AF) çoğunlukla nazofarenks ve posterior nazal kaviteden orijin alırlar. Baş ve boyunun tüm neoplazmları içinde bu lezyonların oranı %0.5'ten daha azdır. Anjiyofibromlar nazofarenks dışında nadirdirler ve bu nedenle yanlış tanı konabilmektedir. Nazal septum oldukça nadir bir bölgedir. Nazal septumdan kaynaklanan ekstra nazofarengal anjiyofibrom şimdiye kadar sadece dört olguda bildirilmiştir. Bu yazıda 19 yaşındaki erkek hastada, nazal septumdan kaynaklanan ve endonazal submukozal rezeksiyon ile çıkarılan, vasküler bir kitle olgusu sunuldu. Bu olgunun histopatolojisi anjiyofibrom olarak bildirildi.

Anahtar Sözcükler: Anjiyofibrom; nazal septum; nazofarengal.

Angiofibromas (AFs) are histologically benign but potentially locally aggressive vascular neoplasms originating characteristically in the posterior lateral wall of the nasopharynx and typically affecting adolescent males.^[1] The incidence of these lesions varies between 0.05 and 0.5% among all neoplasms of the head and neck. They rarely originate from outside the nasopharynx. These unusual tumors are clinically distinct from nasopharyngeal AFs and can therefore be misdiagnosed.^[2] A high level of suspicion is essential for an adequate diagnosis and treatment of these neoplasms.

Our searches of the English language medical literature revealed that only 65 extranasopharyngeal AFs have been reported. The maxillary sinus (24.6-32%) is the commonest site of extranasopharyngeal angiofibromas (ENAs)^[1,3] whereas the nasal septum was an extremely rare site and only four cases of extranasopharyngeal AFs arising from the nasal septum have been reported.^[4-7] Here, we report another case of AF arising from the nasal septum.

CASE REPORT

A 19-year-old male was referred to our hospital with a history of left nasal obstruction and

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recurrent minor nose bleeds for six months. He did not have any history of bleeding disorders. On further examination, a dark purple mass was seen through the left nostril between the nasal septum and the inferior turbinate on anterior rhinoscopy. There was no mass in the nasopharynx on posterior rhinoscopy via the endoscope. The full extent of the mass was not discernible. The right nasal cavity was normal. As it appeared to be a vascular mass it was not probed. There was no cervical lymphadenopathy.

The results of the blood and urine examinations and the chest X-ray were within normal limits. The coagulation profile was normal. The computed tomography (CT) scan demonstrated a tumoral mass confined to the anterior half of the nasal cavity with minimal displacement and erosion of the nasal bones. There was no extension beyond the anterior nasal cavity into the nasopharynx or any other paranasal sinuses (Fig. 1, 2). The nasal septum was deviated to the right side.

In the operation carried out under local anesthesia, it was seen by the endoscope that the mass was connected to the nasal septum. Elevation of the mucoperichondrium was begun through a gently curved vertical incision just cephalic to the caudal end of the septal cartilage. After the elevation of the mucoperichondrium, the septum was visually inspected and the tumor was found to be firmly adherent with the nasal septum at

the junction of cartilage and bone. The mucoperichondrium could not be separated from the tumor. The tumor was removed with a part of the underlying septum. No significant bleeding was observed after surgery. Anterior nasal packing was done with antibiotic embedded gauze. The pack was removed after 48 hours. Postoperative recovery was uneventful. Histopathologic examination showed dilated, cavernous vascular spaces lined by endothelial cells and separated by fibrous tissue and additionally vascular structures covered with monolayer endothelia and fibroblastic cells without atypia and mitosis. Immunohistochemical stains for vimentin exhibited diffuse staining in cells (Fig. 3, 4). Vascular endothelial cytoplasm was stained with CD31. Fibroblastic cells were shown through nuclear and cytoplasmic staining methods through Factor XIIIa. These findings were compatible with the diagnosis of AF. In the five-month follow-up period after the operation, no recurrence has been seen.

DISCUSSION

Angiofibromas arise typically in the nasopharynx, specially at the trifurcation of the sphenoidal process of the palatine bone, the horizontal ala of vomer and the roof of the pterygoid process of the sphenoid at the posterolateral wall of the nasal cavity, near the superior margin of the sphenopalatine foramen. These vascular tumors expand commonly beyond the nasopharynx into the cranium,



Fig. 1. Axial computed tomography scan of the nasal cavity.

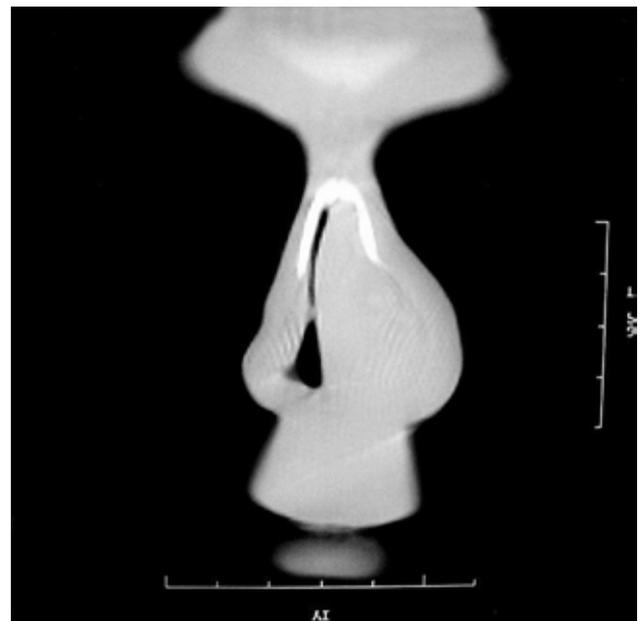


Fig. 2. Coronal computed tomography scan of the nasal cavity.

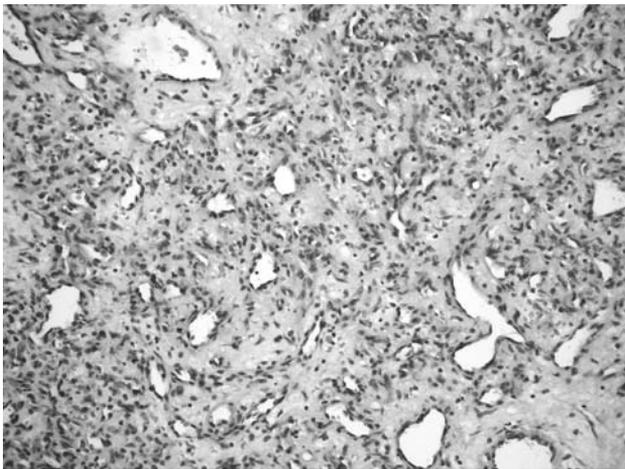


Fig. 3. The histopathological examination of the lesion included vascular structures covered with monolayer endothelia and fibroblastic cells without atypia and mitosis (H-E x 200).

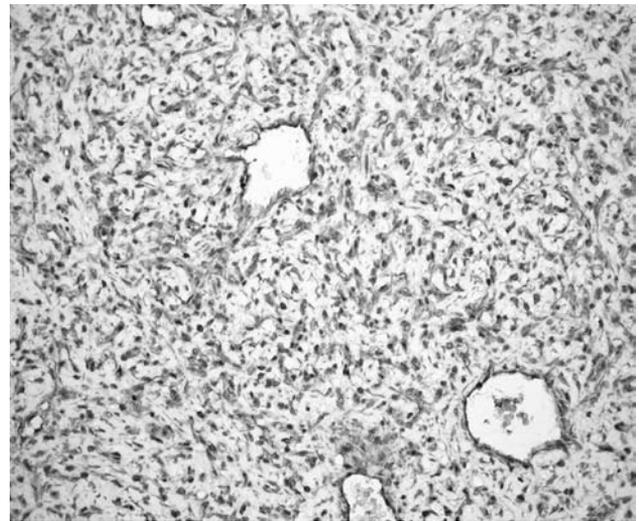


Fig. 4. Immunochemical stains for vimentin exhibited diffuse staining in cells (Vimentin x 100)

nose and paranasal sinuses whereas ENAs by definition arise outside this area. Extranasopharyngeal angiofibroma (ENAs) is rare. The ENAs can originate from any mucosal structure within the head and neck region, including the oral and nasal cavities.^[1,3,8] According to the literature, ENAs are most frequently (32%) localized in the maxillary sinus.^[3] Review of the literature revealed that to date 65 cases have been reported of which only four cases have been arising from the nasal septum.^[1]

By the nature of their location, the presenting symptoms of ENAs are much more variable than NAs: Epistaxis alone or combined with nasal obstruction and facial swelling are the commonest

symptoms of ENAs and the symptoms are developed within 12 months or less before the diagnosis is established.^[4-7] Windfuhr and Remmert^[1] have reported the characteristic age and sex distribution of ENAs: ENAs have a higher mean age, i.e. 22 years as compared to the NA which have a mean age of 17 years. The male to female ratio was 2.75:1. There is a higher incidence of ENA in females. These facts have caused doubt as to whether ENAs -all of them are structurally similar- can be thought of as being different from NAs. Thus, a high level of suspicion is essential for the adequate diagnosis and treatment of these neoplasms. Optimal management of tumors in the

Table 1. Cases of extranasopharyngeal angiofibroma originating from the nasal septum in the literature

Authors	Year	Age/sex	Location	Symptoms	Onset	Therapy
Hiraide and Matsubara ^[4]	1984	13/M	Left nasal septum	Epistaxis, nasal obstruction	2 months	Transnasal resection
Sarpa and Novelty ^[5]	1995	9/M	Right nasal septum	Epistaxis, nasal obstruction	6 weeks	Inspection, manipulation (bleeding); CT, arteriography (no blush)
Handa et al. ^[6]	2001	8/M	Nasal septum	Epistaxis, nasal obstruction	5 months	CT, endonasal removal (bleeding); Open resection
Somdas et al. ^[7]	2005	27/M	Right nasal septum	Epistaxis, nasal obstruction	6 years	–
Present case	2007	19/M	Left nasal septum	Epistaxis, nasal obstruction	6 months	Transnasal resection

CT: Computed tomography.

head and neck area always requires preoperative radiological examination and biopsy. Total surgical resection is the goal of treatment.

Another confusing issue on ENAs is the differential diagnosis of these from other vascular neoplasms. Especially, a dense acellular stroma and excessive collagen tissue is pathognomonic. The size of the vessels may vary from capillaries to venules. In the light of the literature, the most important criterion for differential diagnosis of AFs from other vascular tumors is a dense stromal fibrosis.^[9] The histopathological examination of the lesion confirmed the angiofibroma.

Extranasopharyngeal angiofibroma arising in the nasal septum may be operated, endoscopically or otherwise, through endonasal access; i.e. by lateral rhinotomy, or an alar crease incision (Table 1).

In general, the prognosis for NA is good, although the recurrence rates of 50-61% have been reported. Most of the recurrences are symptomatic within the first year after treatment. Vascularity, age of onset, duration of symptoms and the site and extension of NAs are factors influencing the recurrence.^[10] In contrast to these findings, recurrence was not reported in patients with ENA.

In conclusion, it must be kept in mind that AFs originate from the extranasopharyngeal regions of the head and neck. They must be included in the

differential diagnosis of vascular extranasopharyngeal masses. Angiofibromas are characterized by the predominance of the vascular component in a fibrous stroma which contributes to excessive bleeding following biopsies.

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