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Angiofibroma of the Nasal Vestibule in a 56 Year Old Woman

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Juvenile angiofibroma is a locally aggressive, histologically benign vascular neoplasm seen almost exclusively in males aged 10–25 years. Pathogenetically, the male predilection of the disease suggests a primary aberration of the pituitary gonadal axis. Clinically, angiofibromas appear as well-circumscribed lobulated purple-red masses covered by intact mucosa on the nasopharyngeal surface. Histologic study reveals two main cellular components: a fibrous stroma, and a rich network of irregularly shaped blood vessels. The vessels vary from very small endothelium-lined capillaries to large venous channels. They lack the smooth muscle and elastic fibers found in normal blood vessels, such that even minimal manipulations can lead to hemorrhage [1].

Angiofibromas originate in the superior margin of the sphenopalatine foramen. They are slow-growing but locally invasive, and can be life-threatening because of bleeding or intracranial extension. Intracranial extension is reported in 10%–36% of cases [2]. The most common symptom, in about 80% of patients, is unilateral nasal obstruction and epistaxis. Other symptoms and signs are facial swelling, proptosis, and ocular problems. Computed tomography and magnetic resonance imaging serve as diagnostic tools and are also useful for pre- and postoperative evaluation.

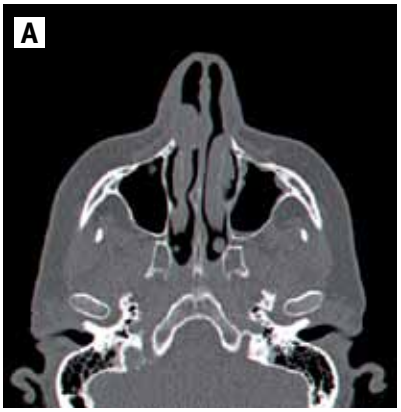
Surgery is the treatment of choice, although radiation therapy had been used with reasonable efficacy. The surgical approach is determined primarily by the location and extent of the tumor. Many traditional approaches allow good access for resection, but not without bony and soft tissue dissection. Transnasal endoscopic resection, first reported in 1996, has been found to be a suitable alternative to surgery [3]. Recurrence is a predominant feature of angiofibroma, with rates ranging from 30% to 46% [4]. In the present report we describe an unusual case of juvenile angiofibroma in a middle-aged woman.

PATIENT DESCRIPTION

A 56 year old woman, with an unremarkable history, presented to our clinic with a slow-growing swelling of the right nasal vestibule of 3 years duration. She had no obstructive complaints, and no epistaxis was noted. Physical examination revealed a purple-red mass in the right nasal vestibule measuring 3 cm in diameter. The mass was covered anteriorly by skin and posteriorly by mucosa. There was no bleeding. CT scan showed a 2 x 2 x 3 cm round mass located in the right nasal vestibule, causing complete nasal obstruction [Figure A], without displacement of the nasal septum or turbinates.

Following premedication with intramuscular pethidine and atropine, local nasal anesthesia consisting of a mix of adrenaline, bupivacaine, and lidocaine 2% was applied. A purple-red obstructing mass was seen in the right nasal vestibule via a 0-degree endoscope. The mass was not attached to the nasal septum but to the inferior concha. En bloc resection was performed, leaving the nasal airway completely open. Bleeding was controlled with the insertion of anterior nasal packs.

[A] Sagital CT scan demonstrating a soft tissue mass occupying the right nasal vestibule area

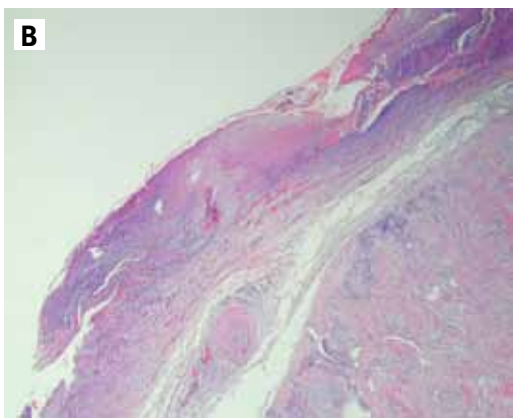


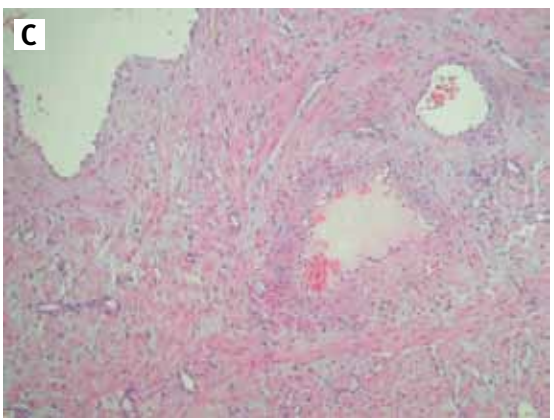
On the second postoperative day, the anterior packing was removed. There was no bleeding or infection. Two months later, the nose was healed completely, without any residual disease.

Macroscopic examination showed a solid grey-white nodular mass measuring 2 cm in diameter. Microscopically, a tissue fragment covered by benign squamous epithelium was noted on one side of the specimen [Figure B]. Beneath the epithelium was a well-circumscribed nodular mass composed of numerous, variably sized, endothelium-lined vascular channels. Fibroblasts, fibrocollagenous elements and spindle-shaped smooth muscle cells were interspersed among the channels [Figure C].This was consistent with angiofibroma. There was no evidence of malignancy.

COMMENT

We describe a 56 year old woman with an angiofibroma originating in the right





[B] Well-circumscribed nodular mass in which one surface is covered by benign stratified squamous epithelium (x40)
[C] Stroma showing endothelium-lined vascular channels interspersed by fibroblasts, collagen, and spindle-shaped smooth muscle cells (x100)

nasal vestibule area. The present case is unique for the patient's female gender, which has been reported only rarely in angiofibroma [5], older age, and origin of the lesion far from the traditional location at the superior margin of the sphenopalatine foramen. Because our patient's background features were not compatible with angiofibroma, she was not referred for angiography or embolization prior to surgery.

To conclude, this case should alert physicians to the possibility of exceptional occurrences of angiofibroma, warranting a high index of suspicion.

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