Retropharyngeal Angiofibroma

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Juvenile nasopharyngeal angiofibromas (JNAs) frequently occur in young males and originate from the lateral wall of the nasopharynx. JNAs are highly vascularized and histologically benign tumours with locally aggressive behaviour. These tumours may be associated with significant mortality and morbidity due to their local destruction and vascularity. Major symptoms are epistaxis and nasal obstruction. The incidence of these tumours is 1:5000 to 1:60 000, and they account for only 0.5% of all head and neck neoplasms.1

JNA is very rare in individuals over the age of 25, in female patients, and outside the nasopharyngeal area. To date, approximately 60 extranasopharyngeal angiofibromas have been reported in the English literature.2 Histologically, an angiofibroma is defined as a noncapsulated tumour composed of blood vessels and fibrous connective tissue. Favoured treatment is by surgical excision of the mass. In this case report, a middle-aged female patient with angiofibroma located in the retropharyngeal space is presented.

Case Report

A 35-year-old female patient was admitted to our clinic with the complaints of difficulty on swallowing, hoarseness, and aphony for 3 years. On oral examination, a smooth-surfaced, pinkish purple, round mass originating from the posterior wall of the pharynx was observed. On direct laryngoscopy, a bulging behind the posterior pharyngeal wall extending toward the cricoid cartilage level was evident (Figure 1). On magnetic resonance imaging (MRI), a 72 × 47 × 30 mm solid round mass with regular borders originating from the posterior wall of the pharynx and extending toward the epiglottis was observed (Figure 2). The blood supply of the mass was shown by angiography to be mainly from the lingual branch of the external carotid artery on the right side, and the mass was fed by the internal maxillary and lingual branches of the external carotid artery on the left side (Figure 3).

We decided to proceed with surgical excision of the mass. To expose the mass, an apron incision was used. During surgery, the retropharyngeal mass was seen extending from the oropharyngeal region superiorly toward the postcricoid area inferiorly (Figure 4). The tumour was totally excised by working on both sides of the larynx, without injuring the pharyngeal mucosa or the recurrent laryngeal nerves. A tracheostomy was performed for airway safety. The excised specimen was 7 × 5 × 3 cm in size and had lobulated contours (Figure 5). The histopathologic examination was reported as angiofibroma (Figure 6). The tracheostomy tube was removed on the seventh day following the procedure. Follow-up at 18 months revealed the patient to be free of disease, without any complaints.

Figure 1. Appearance of the mass on oropharyngeal examination.
Discussion

Nasopharyngeal angiofibromas almost always occur in men, although the female to male ratio for extranasopharyngeal angiofibromas is 1:3. The mean age at occurrence is 17 years for nasopharyngeal and 22 years for extranasopharyngeal angiofibromas. Nasopharyngeal angiofibromas are associated with symptoms of nasal obstruction, pain, and epistaxis; however, symptoms due to extranasopharyngeal angiofibromas vary by the area of origin. The clinical appearance of an angiofibroma is a firm mass with a pinkish purple colour. Local spread of the tumour can cause significant mortality and morbidity, especially if it is highly vascular or there is intracranial extension. The vascular component is dominant in angiofibromas, and punch biopsy of the mass can cause excessive bleeding and therefore is contraindicated. Angiofibromas are rarely seen outside the nasopharyngeal area, but maxillary, ethmoid, sphenoid, nasal septum, middle and inferior turbinate, laryngeal, and conjunctival locations have been reported. The most common site of origin of an extranasopharyngeal tumour is in the maxillary sinus. Our case is a very atypical one because of its retropharyngeal origin and the patient’s age and sex. Our review of the literature revealed only two cases of laryngeal angiofibroma, but a retropharyngeal location has never been reported.

The most commonly employed radiologic modalities are computed tomography, MRI, magnetic resonance angiography, and selective angiography of the external and internal carotid arteries. MRI is a useful technique for the evaluation of soft tissue mass lesions of the retropharyngeal and hypopharyngeal areas. The size, extension localization, vascularization of the mass, and involvement of the surrounding bony structures may be determined.

The definitive diagnosis of angiofibromas is by histopathologic analysis of the surgical specimen. On microscopic examination, many irregular blood vessels of various sizes, lined with one layer of endothelium, were seen in a fibrocollagenous stroma. The large vessels contained an interrupted medial layer. Vascularity was more prominent in the periphery of the tumour and consisted of small vessels. The stroma between the vessels consisted of spindle-shaped and stellate fibroblastic cells that were scattered in an irregular, collagenous matrix.

Figure 2. Magnetic resonance image revealing a contrast-enhancing mass in the retropharyngeal area.

Figure 3. Angiogram showing a highly vascular mass supplied by the lingual branch of the external carotid artery on the right side (A) and by the internal maxillary and lingual branches of the external carotid artery on the left side (B).
There were no mitoses. The histopathologic diagnosis in light of these findings was angiofibroma.

The theories of origin and histogenesis of angiofibroma are varied (developmental, hormonal imbalance, and genetic causes). According to Tillaux, these tumours originate from the tissue at the anterior margin of the atlas, at the lower surface of the sphenoid bone, known as the fibrocartilaginous barrier.\(^5\) Brunner called this tissue “fascia basalis” because he found no cartilage in it.\(^{11,12}\) Hiraide and Matsubara reported cases located at the anterior third of the nasal septum, at the junction of the cartilage, and the perpendicular plate of the ethmoid bone, which led them to suggest that this tumour arises from the periosteum of the perpendicular plate of the ethmoid bone where the fascia basalis exists.\(^{12}\) Conversely, there is no fascia basalis in the nasal septum except in the posterior part of the vomer and the ethmoid bone.\(^{11}\) Because there is no fascia basalis in this region, the site of origin of extranasopharyngeal angiofibroma is debatable. Following these observations, it can be concluded that angiofibromas arise from an ectopic tissue and may be related to developmental anomalies. The location of our case supports this theory. According to Schiff, it is possible that turbinate-like vascular tissue is retained as an ectopic nidus in developing periosteum during the growth of the nasal septum. Hora and Weller reported a maxillary sinus angiofibroma associated with maxillary deformity and congenital cleft palate and lip.\(^{13}\)

**Conclusion**

It must be kept in mind that angiofibromas can originate in the extranasopharyngeal regions of the head and neck. They must be included in the differential diagnosis of vascular masses.

**Acknowledgement**

Financial disclosure of authors and reviewers: None reported.

**References**


