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Hypervascular Tumor of the Buccal Space in an Adult as a Late Recurrence of Juvenile Angiofibroma

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Summary: We describe an adult patient with recurrent juvenile angiofibroma, which presented as a rapidly enlarging, hypervascular mass in the anterior part of the cheek. The case is unusual because of the extreme delay (greater than 30 years) and the anatomic location of the recurrence.

Index terms: Angiofibroma; Face, neoplasms

Juvenile angiofibromas are relatively rare fibroangiomatous tumors of the nasopharynx, posterior nasal cavity region. Typically, these tumors develop in adolescent boys, although there have been occasional reports of juvenile angiofibromas occurring de novo in adult men and women (1–3) and in girls (4).

Although histologically benign, these tumors are usually locally aggressive, frequently extending well beyond the nasopharynx along natural tissue planes, foramina, and fissures that interconnect the defined compartments of the head and neck (1). Owing to their predilection for local invasion, optimal surgical management of juvenile angiofibromas can be difficult, contributing to a variably high rate of recurrence. Recurrent disease is usually seen within months to a few years after primary treatment, and commonly involves regions of difficult surgical accessibility to which the tumor has directly extended, such as the intracranial contents and the infratemporal fossa (5).

We report a case of recurrent juvenile angiofibroma in a middle-aged man that presented as a rapidly enlarging mass of the anterior part of the cheek. This case is particularly unusual in that the recurrence developed more than 30 years after the original treatment and was centered within the buccal space. This unusual recurrence of juvenile angiofibroma in an adult illustrates the potential of these tumors to recur in unusual locations after long periods of biological quiescence.

Case Report

A 44-year-old man presented with a left-sided facial mass that had rapidly enlarged over a 7-month period. His medical history was notable for nasopharyngeal juvenile angiofibroma at age 10 years, which had been treated with presumably incomplete resection and local radiation therapy. Details of the patient's previous treatment were unknown owing to the unavailability of his medical records.

On physical examination, there was a large oval mass in the anterior part of the lower left cheek, extending posteriorly to the coronoid process. The overlying skin was erythematous. No lymph nodes were palpated.

Contrast-enhanced computed tomography (CT) of the face (Fig 1A) showed a 6-cm lobular mass centered within the left buccal space with extension into the adjacent masticator space posteriorly and superiorly within the infratemporal fossa. There was associated mild bony remodeling of the lateral wall of the left maxillary sinus. The mass showed prominent inhomogeneous contrast enhancement with central areas of low attenuation that were thought to represent necrosis.

Magnetic resonance (MR) imaging (Fig 1B) showed the mass to be of intermediate signal intensity on T1-weighted images and of high signal intensity on T2-weighted images. The epicenter and extent of the tumor were the same as noted by CT. The mass enhanced intensely and uniformly after administration of gadopentetate dimeglumine. An attempt was made at transbuccal biopsy and fine-needle aspiration, but these were nondiagnostic and yielded only bright red blood from the mass.

Selective left external carotid angiography (Fig 1C) showed intense neovascular blush of the tumor with predominant supply from buccal and nasal branches of the left distal internal maxillary artery. Major supply was also seen arising from the transverse facial artery, and minor supply to the tumor was noted from the ascending palatine
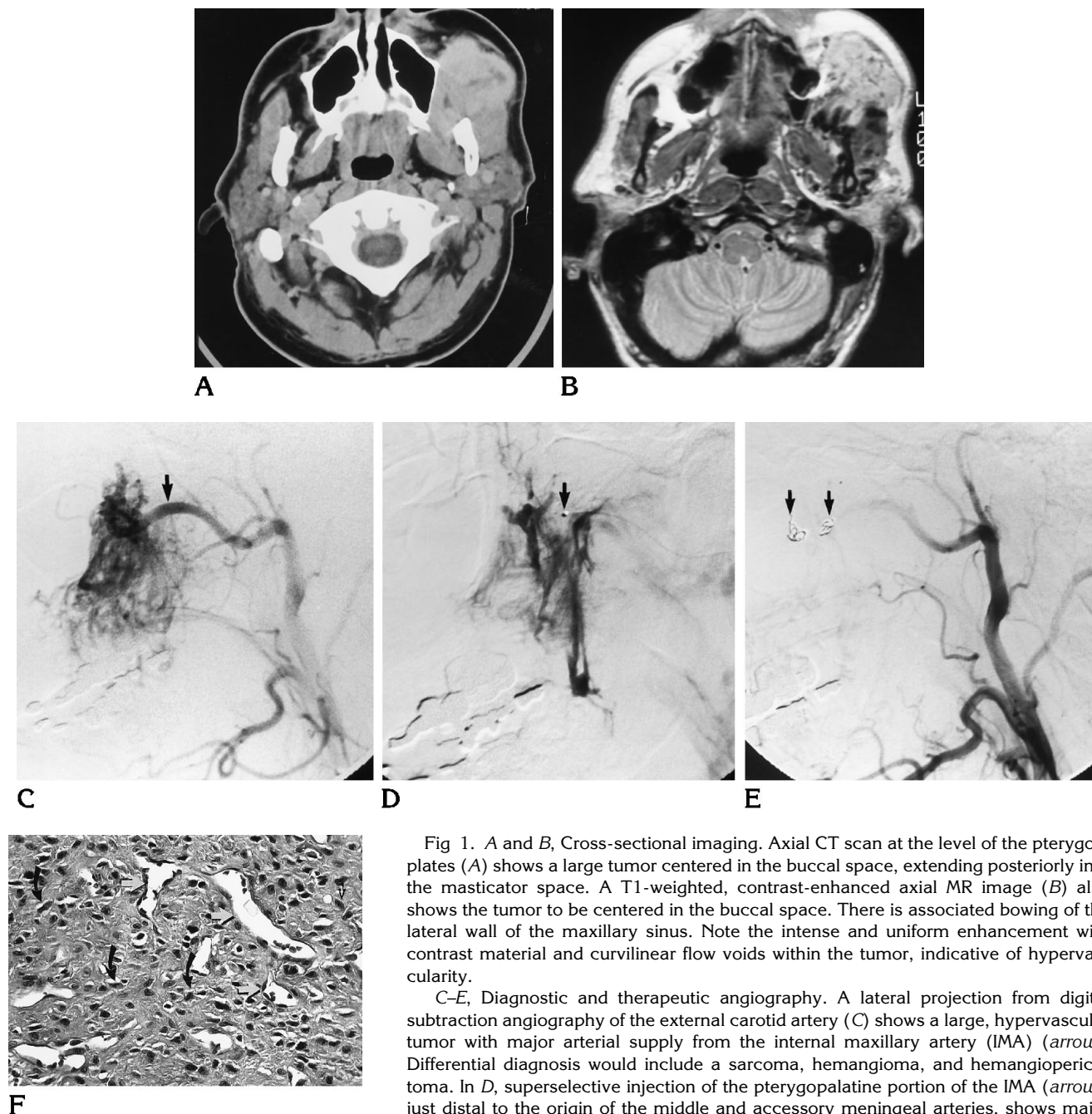
Fig 1. *A* and *B*, Cross-sectional imaging. Axial CT scan at the level of the pterygoid plates (*A*) shows a large tumor centered in the buccal space, extending posteriorly into the masticator space. A T1-weighted, contrast-enhanced axial MR image (*B*) also shows the tumor to be centered in the buccal space. There is associated bowing of the lateral wall of the maxillary sinus. Note the intense and uniform enhancement with contrast material and curvilinear flow voids within the tumor, indicative of hypervascularity.

C–E, Diagnostic and therapeutic angiography. A lateral projection from digital subtraction angiography of the external carotid artery (*C*) shows a large, hypervascular tumor with major arterial supply from the internal maxillary artery (IMA) (*arrow*). Differential diagnosis would include a sarcoma, hemangioma, and hemangiopericytoma. In *D*, superselective injection of the pterygopalatine portion of the IMA (*arrow*), just distal to the origin of the middle and accessory meningeal arteries, shows major arterial supply to the tumor. Particulate embolization was performed with the microcatheter in this position. A final external carotid angiogram (*E*) shows near-total devascularization of the tumor after both particulate and coil embolization of the distal IMA (*arrows*).

F, Histologic section shows that the tumor consists of fibrovascular tissue with variable hypecellularity and fiber density. There are numerous plump spindle cells (*curved arrows*) and mixed inflammatory cells. Note the many ectatic, thin-walled vascular channels (*arrows*) (hematoxylin-eosin stain; magnification ×350).

branch of the left facial artery. Selective left internal carotid injection showed minor supply to the tumor from the artery of the foramen rotundum. No contralateral carotid supply to the tumor was seen.

Owing to the extensive hypervascularity of the tumor, preoperative therapeutic embolization was performed. A guiding catheter was positioned in the proximal left external carotid artery, and microcatheters were used to superselectively catheterize the major feeding pedicles to the tumor (ie, the distal internal maxillary and transverse facial arteries). These pedicles were embolized serially with suspensions of polyvinyl alcohol particles (Fig 1D). A final

common carotid angiogram (Fig 1E) showed greater than 90% of the tumor supply had been occluded with minimal persistent supply from the artery of the foramen rotundum.

Two days later, the juvenile angiofibroma was completely resected via a transbuccal and lateral maxillectomy approach. Pathohistologic examination of the resected specimen showed fibrovascular tissue with varying degrees of fiber density and cellularity, thin-walled ectatic vessels, and a mixture of inflammatory cells and plump, focally atypical spindle cells (Fig 1F). These findings were compatible with a pathohistologic diagnosis of recurrent juvenile angiofibroma.

Discussion

Although juvenile angiofibromas classically develop in adolescent boys, they occasionally may occur in infants, adults, and girls (1, 4, 5). The specific origin of these tumors remains speculative; theories of pathogenesis include fibroangiomatous hyperplasia (6), hamartomatous ectopia (7), and paraganglionic hyperplasia (8). There is immunohistochemical evidence to suggest that juvenile angiofibromas have abundant noradrenergic innervation (9) and appear to contain an angiogenic growth factor (10).

Originally it was believed that juvenile angiofibromas arise within the nasopharynx; however, it is currently theorized that these tumors arise along the posterolateral wall of the nasal cavity, where the sphenoidal process of the palatine bone meets the horizontal ala of the vomer and the root of the pterygoid process. This area includes the superior margin of the sphenopalatine foramen and the posterior end of the middle turbinate, thus explaining the ease with which the tumor spreads to the sphenoidal sinus, nasopharynx, pterygopalatine fossa, pterygomaxillary fissure, and infratemporal fossa. There have also been occasional reports of juvenile angiofibromas manifesting as a primary tumor within the maxillary sinus (2, 11). These observations have prompted some authors to avoid the term *nasopharyngeal* when referring to these tumors.

Despite their characteristically benign histopathologic appearance by light microscopy, juvenile angiofibromas tend to be locally aggressive tumors that have a propensity to extend some distance from their primary site of origin. These tumors typically begin to spread by submucosal extension into the nasal cavity, posterior nasopharynx, pterygopalatine fossa, and sphenoidal sinus (1, 5). Lateral extension from the pterygopalatine fossa into the pterygomaxillary fissure and infratemporal fossa is also frequent, whereas intraorbital and superior intracranial invasion is less frequently observed but not uncommon (5).

As a consequence of their local aggressiveness, optimal surgical resection of juvenile angiofibromas is often challenging, which most likely substantially contributes to their variably high rate of recurrence after first attempts at surgical management (perhaps as high as 60% in some series). However, good control rates (up to 90%) have been reported in the treatment of recurrences by either surgery or radiation therapy (1, 5, 12). Recurrent disease is usually seen within the first 3 years, and most commonly involves regions that are difficult to access surgically, such as the intracranial contents and the infratemporal fossa (5).

Surgical resection is generally believed to be the treatment of choice, although some have advocated the use of radiation therapy as the preferred method for advanced juvenile angiofibromas (particularly when seen in association with intracranial extension) (13). With the advent of multidisciplinary teams specializing in surgery of the cranial base, many centers are now advocating surgery as the primary approach to advanced juvenile angiofibromas (14). Radiation therapy has fallen into some disfavor owing to a combination of improved surgical techniques and the combined risk of sarcomatous degeneration of irradiated juvenile angiofibromas and the development of radiation-induced second primary neoplasms (15–17).

In the past, an additional major problem of definitive surgical resection of juvenile angiofibromas has been the large, sometimes fatal, loss of blood during extirpation (1). To overcome this problem, endovascular therapeutic devascularization has been widely adopted as an important preoperative therapeutic maneuver (18–20). Transarterial embolization through superselective catheterization of supplying arterial pedicles with polyvinyl alcohol particles remains the standard conventional endovascular therapeutic approach to these tumors, although recently, direct puncture techniques have been used successfully, with liquid adhesives injected directly into the tumor neovasculature (19).

In our patient, we speculate that the juvenile angiofibroma was originally incompletely resected, in which a laterally extending tongue of

tumor was left in the lower portion of the ptery-gomaxillary fissure. This residual tumor eventually spread directly into the masticator space of the infratemporal fossa and subsequently inferiorly and anteriorly into the buccal space, where the bulk of the tumor was located. It also is possible that this inferior extent of the juvenile angiofibroma was not included within a radiation port, further increasing the chance of recurrence, although records of the patient's original treatment are no longer available to substantiate this theory.

Frequently, recurrent disease manifests as a lateral cheek mass caused by extension from the pterygomaxillary fissure into the masticator space (1, 5). In such situations, the bulk of the tumor is centered within the masticator space with minor secondary involvement occasionally seen in the buccal space. We cannot offer an explanation as to why our patient had a diametric distribution of recurrent tumor.

The extremely long period of biological quiescence of our patient's juvenile angiofibroma is also exceptional, since most clinical series have reported recurrences developing within months to a few years after primary treatment (1, 5, 12). Longer periods of latency have occasionally been reported; all in association with the development of either malignant sarcomatous degeneration or a second primary tumor caused by radiation therapy (fibrosarcoma and malignant fibrous histiocytoma) (15, 16, 17). In these previous reports, time intervals from treatment to malignant recurrence ranged from 5 to 18 years. Although this possible complication of therapy was initially strongly suspected in our patient, histologic examination of the resected specimen was most consistent with a simple recurrence of juvenile angiofibroma.

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