



CASE REPORT

Buccal Space Giant Cell Angiofibroma: Case Report and Review of Head and Neck Occurrences

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Abstract

Background: Giant cell angiofibroma (GCA) was first described in 1995, as a previously unrecognized soft tissue tumor of the orbit. Since that time, rare extraorbital head and neck manifestations of GCA have been documented. To the best of our knowledge, we present the fourth documented case of GCA of the buccal cavity, constituting the largest dominant site of occurrence in the extraorbital head and neck.

Methods: This is a single-surgeon retrospective case report and review of the literature of patients with extraorbital head and neck manifestations of giant cell angiofibroma.

Results: There have been 8 extraorbital head and neck manifestations of giant cell angiofibroma, 4 within the buccal cavity. GCA occurring outside the head and neck, seems to occur more often in females. On the contrary, head and neck manifestations seem to have a male predilection. Extraorbital head and neck manifestations of GCA occurs primarily in adulthood at a median age of 45 (range 29-61), which has a similar distribution to orbital and non-head and neck sites. The highest age distribution seems to be in the fifth decade of life.

Conclusions: Extraorbital head and neck manifestation of GCA represents a unique disease entity with a distinctive gender predilection different from that of other sites. In the review of the literature, it seems that the buccal mucosa is the most prevalent extraorbital head and neck site. Awareness of the lesion, and its close relationship with other soft tissue tumors is important in the differential diagnosis and management.

Keywords

Giant cell angiofibroma, Rare head and neck tumor, Head and neck surgery

Introduction

Giant cell angiofibroma (GCA) was first described by Dei Tos, et al. in 1995, as a previously unrecognized soft tissue tumor of the orbit [1]. It was pathological characterized as having patternless round to spindle cell proliferation with moderate to high degree of cellularity. All cases further exhibited prominent vascularity, vessel wall hyalinization, and lacunar and irregularly shaped pseudovascular spaces containing granular material. Since that time, rare extraorbital head and neck manifestations of GCA were documented in the scalp, vocal cord, oral cavity, submandibular region, and parapharyngeal space [2-6]. To the best of our knowledge, we present the fourth documented case of GCA of the buccal cavity, constituting the largest dominant site of occurrence in the extraorbital head and neck.

Case Report

A 29-year-old otherwise healthy African American male presented for evaluation of a right buccal mass present for 1 year. Based on patient history, the mass had exhibited slow, painless growth. The mass was located within the right buccal mucosa, measuring approximately 2.5 × 2.0 cm, approximately 2 cm posterior to the oral commissure. On exam, it was soft, freely mobile, non-fluctuant, without changes to the overlying mucosa. The patient underwent a transoral resection of the mass, with primary closure. Intraoperatively, a transoral incision was made just 1 cm anterior to lesion, and

through blunt dissection the lesion was easily delivered from the buccal space. The patient was closed primarily, without any postoperative complications.

Microscopical analysis revealed a well delineated neoplasm composed of proliferating bland spindle cells with intervening collagen deposition and foci of giant cell formation. Irregular capillaries were also present. The neoplasm was found to be bcl-2, CD34, CD99, and Factor XIIIa positive, and s-100 and smooth muscle actin negative.

Discussion

Giant cell angiofibroma was originally described as a unique orbital tumor by Dei Tos, et al. in a series of seven patients, of whom 6 were male, with a median age

of 59 (range 24-73) [1]. It was thought only to occur in the orbit, and as such the original authors designated the lesion "Giant Cell Angiofibroma of the Orbit". All lesions were found in the eyelid, in the region close, or involving the lacrimal gland. Within their study they described the lesion as sharing similar characteristics but distinct from solitary fibrous tumors and giant cell fibroblastoma.

Since its original description, rare cases have been described in approximately 18 extraorbital location including the mediastinum, parascapular region, retroauricular sites, thigh, retroperitoneum, vulva, hip, forearm, groin [2,7]. Within the head and neck, extraorbital cases have been documented in the scalp, vocal cord, oral cavity, submandibular region, and parapharyngeal spaces (Table 1).

Table 1: Literature review of documented giant cell angiofibromas in the head and neck with pathologic description.

Authors	Head and Neck Site	No. Cases	Age Range	Gender Distribution	Histopathologic Features	Immunohistochemical Staining
Arifin, et al. [2]	Scalp	1	39	Female	Oval and round- to spindle-shaped cells intermixed with floret-like giant with hypocellular areas. Giant cells lined the pseudovascular space. Presence of bony erosion.	Positive: CD34 and Vimentin Negative: not mentioned
Shu, et al. [3]	Vocal cord	2	40-45	All Male	Patternless spindle cells with rich blood vessels and vascularity, with presence of multinucleated cells lining pseudovascular spaces. Positive training: CD34, Vimentin, CD99. Negative staining.	Positive: CD34, vimentin, CD99 Negative: MSA, desmin, s100, aAT, aACT, CDC9, cytokeratin
Gonzalez-Perez, et al. [4]	Parapharyngeal Space	1	29	Female	Fibrous pseudocapsule of spindle cells, round to oval bland nuclei and pseudoinclusions. Numerous floret like multinucleated giant cells, angioectoid pseudovascular spaces.	Positive: CD34, BCL2, CD99 Negative: smooth muscle actin, CKAE1/AE3, CK1/5/10/14, s100, p63, CD31, EMA <i>In-situ</i> hybridization negative to SYT gene (18q11.2) to rule out synovial sarcoma
Rousseau, et al. [5]	Buccal Mucosa	1	61	Male	Presence of torturous, pseudovascular spaces, filled with floret type multinucleate cells with large hyperchromatic nuclei.	Positive: Vimentin, CD34 Negative: CD68, CD31, Factor VIII, <i>Ulex europaeus</i> lectin
De Andrade, et al. [9]	Buccal Mucosa	1	44	Male	Elongated, spindled, and wavy cells, intermingled with variable amounts of blood vessels, pseudovascular spaces, and scarce multinucleated giant cells.	Positive: Vimentin, CD34, bcl-2 (weakly) Negative: cytokeratin, smooth muscle actin, s100, desmin, CD68, CD34
Kintarak, et al. [8]	Buccal Mucosa	1	46	Female	Nonencapsulated, well circumscribed, with small spindle shaped proliferating cells. Scattered multinucleated giant cells with peripherally arranged nuclei in floret like pattern, lining pseudovascular spaces.	Positive: CD34, factor XIIIa Negative: smooth muscle actin, desmin, s100
Mikami, et al. [6]	Submandibular Region	1	48	Female	Pseudovascular spaces lined by a discontinuous row of multinucleated cells were seen against a background of spindle-shaped fibroblastic cell proliferation.	Positive: CD34, Vimentin Negative: Factor VIII, desmin, smooth muscle actin, myoglobin, s100, leuM1, lysozyme, alpha-1-antitrypsin, cytokeratin

In review of the case report literature, it was found that there is no gender predilection in extraorbital head and neck sites, in contrast to the original orbital lesion found mostly in males. Furthermore, GCA occurring outside the head and neck, in regions such as the thigh, retroperitoneum, hip and forearm, seems to occur more often in females [9,10]. Extraorbital head and neck manifestations of GCA occurs primarily in adulthood at a median age of 45 (range 29-61), which has a similar distribution to orbital and non-head and neck sites. The highest age distribution seems to be in the fifth decade of life.

Pathologic assessment of GCA from the extraorbital head and neck uniformly reveals an unencapsulated, well circumscribed lesion with the presence of round to spindle shaped cells and giant cells lining pseudovascular spaces. All lesions are benign appearing, with an indolent course, treated with whole specimen surgical excision, without documentation of recurrence. However, one documented case by Arifin, et al. revealed the presence of bony erosion on pathologic assessment and radiographic imaging of a lytic lesion [2]. However, despite the bony erosive changes, no documentation of malignant behavior or recurrence was evident. Furthermore, GCA lesions uniformly stain positive for CD34 and vimentin, with a no staining to cytokeratin, smooth muscle actin, s100, demisin, p63, CD31. Gonzalez-Perez, et al. further performed *in situ* hybridization studies to rule out synovial sarcoma, with probes to the SYT gene (18q11.2) that were determined to be negative [4].

Given the rarity of GCA in the extraorbital head and neck, the authors describe a tumor that has a propensity to occur in the soft tissue of the head and neck. In the review of the literature, it seems that the buccal mucosa is the most prevalent extraorbital head and neck site. Awareness of the lesion, and its close relationship with other soft tissue tumors is important in the differential diagnosis and management.

Conflict of Interests

None.

Financial Disclosures

None.

Competing Interests

None.

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