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ORIGINAL PAPER

Angioleiomyoma of the Sinonasal Tract: Analysis of 16 Cases and Review of the Literature

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Abstract Angioleiomyoma (ALM; synonyms: angiomyoma, vascular leiomyoma) is an uncommon benign tumor of skin and subcutaneous tissue. Most arise in the extremities (90 %). Head and neck ALMs are uncommon (~10 % of all ALMs) and those arising beneath the sinonasal tract mucosa are very rare (<1 %) with 38 cases reported so far. We herein analyzed 16 cases identified from our routine and consultation files. Patients included seven females and nine males aged 25–82 years (mean 58; median 62). Symptoms were intermittent nasal obstruction, sinusitis, recurrent epistaxis, and a slow-growing mass. Fifteen lesions originated within different regions of the nasal cavity and one lesion was detected incidentally in an ethmoid sinus sample. Size range was 6–25 mm (mean 11). Histologically, all lesions were well circumscribed but non-encapsulated and most (12/16) were of the compact solid type superficially mimicking conventional leiomyoma but contained numerous compressed muscular veins. The remainder were of venous (2) and cavernous (2) type. Variable amounts of mature fat were observed in four cases (25 %). Atypia, necrosis, and mitotic activity were absent. Immunohistochemistry showed consistent expression of smooth muscle actin (12/12), h-caldesmon (9/9), muscle-specific actin (4/4), variable expression of desmin (11/14) and CD56 (4/6), and absence of HMB45 expression (0/11). The covering mucosa was ulcerated in 6 cases and showed squamous metaplasia in one case. There were no recurrences after local excision. Submucosal sinonasal ALMs are rare benign tumors similar to their reported cutaneous counterparts with frequent adipocytic differentiation. They should be distinguished from renal-type angiomyolipoma. Simple excision is curative.

Keywords Angioleiomyoma · Sinonasal tract · Angiomyolipoma · Vascular leiomyoma · Angiomyoma · PEComa · Nasal

Introduction

Mesenchymal tumors of the sinonasal tract are rare. They encompass benign tumors (benign peripheral nerve sheath tumors, angioleiomyoma and hemangiomas), lesions of low-grade or uncertain biological potential (sinonasal hemangio/glomangiopericytoma, solitary fibrous tumor, desmoid fibromatosis, low-grade malignant peripheral nerve sheath tumors and low-grade sinonasal sarcoma with neural and myogenic features) and frankly malignant aggressive neoplasms (conventional malignant peripheral nerve sheath tumors, leiomyosarcoma, rhabdomyosarcoma and other rare sarcoma types) [1–5]. Due to the rarity of sinonasal mesenchymal neoplasms, many pathologists are not familiar with their broad phenotypic spectrum.

Angioleiomyoma (ALM; synonyms: angiomyoma, vascular leiomyoma) is an uncommon benign tumor of skin and subcutaneous tissue composed of well differentiated smooth muscle proliferations associated with variable but usually prominent vascular component [6]. The latter may consist of thick-walled collapsed vascular channels (solid type), predominant thick-walled venous vessels with well

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recognizable lumens within smooth muscle background (venous type), or display ectatic muscular venous channels mimicking venous hemangioma but with variable smooth muscle component in-between (cavernous type) [6]. The majority of lesions originate in the extremities (~90 %), mainly in the lower limbs while ALM of the head and neck region is uncommon (~10 %) [7, 8]. Submucosal ALMs of the sinonasal tract are exceptionally rare. To date, 38 cases have been reported, mostly as single case reports [9–41] (Table 1). In this study, we describe our experience with submucosal ALMs of sinonasal tract and discuss their clinicopathological and immunohistochemical characteristics in light of previously reported cases with special emphasis on the frequent presence of adipocytic differentiation and similarities and differences compared to their cutaneous and soft tissue counterparts.

Materials and Methods

We reviewed our routine and consultation files for lesions coded as angioleiomyoma, angiomyoma, angiomyolipoma and vascular leiomyoma originating in the nose, nasal cavity, or paranasal sinuses. Diagnosis was based on criteria defined for similar lesions in the most recent World Health organization (WHO) classification of tumors of the head and neck and tumors of soft tissue and bone [1, 6]. After review, only tumors arising from the mucosa-lined sinonasal sites (excluding cutaneous lesions) were included in this series. The tumor specimens were fixed in buffered formalin and embedded routinely for light microscopic examination. Immunohistochemical studies were performed on 3–4-μm sections cut from paraffin blocks using a fully automated system ("Benchmark XT System", Ventana Medical Systems Inc, Tucson, Arizona, USA) using the following antibodies: α-smooth muscle actin (clone 1A4, 1:200, Dako), h-caldesmon (clone h-CD, 1:100, Dako), desmin (clone D33, 1:250, Dako), CD34 (clone QBEnd10, 1:200, Immunotech), CD56 (clone MRQ-42, 1:100, CELL MARQUE), HMB45 (clone HMB45, 1:50, Loxo), and podoplanin (clone D2-40, 1:50, Zytomed).

Results

Clinical Features

Sixteen cases were retrieved from our files (Table 1, Cases 39–54). Patients included seven females and nine males aged 25–82 years (mean 58; median 62). Variable combinations of intermittent nasal obstruction, chronic sinusitis-like symptoms and recurrent epistaxis were the presenting symptoms in eight patients with detailed data, respectively. A painful mass was stated in one case. Other cases either lacked detailed history or a slowly growing mass or nodule was the presenting symptom of the disease. The site was stated within different compartments of the nasal cavity (five in turbinates, four in the nasal cavity unspecified, three from the nasal septum, two in the nasal orifices, and one in the lateral nasal wall). One case (the only sinus-based lesion) was found incidentally in a polyposis specimen from the ethmoid sinus. None of the lesions was multifocal or involved more than one subregion of the sinonasal tract. There was no evidence of associated diseases or similar tumors elsewhere in the body. All lesions were removed via simple complete local excision with free albeit close margins.

At last follow-up (range 9–211 months; mean 58 months), no recurrences were recorded.

Pathological Findings

Tumor size ranged from 6 to 25 mm (mean 11) in cases with gross description or as measured from glass slides. Grossly, the lesions were described as tan-whitish with solid whorled cut-surface and firm consistency. Histologically, all lesions were non-encapsulated but well circumscribed (Fig. 1a, b). The tumors were covered by sinonasal mucosa with variable reactive or metaplastic changes (Fig. 1c). Six tumors showed mucosal ulceration/erosion with variable inflammation between the tumor and mucosal surface associated with variable degree of fibromyxoid vascular obliteration (Fig. 1d). The tumors were composed of well differentiated smooth muscle cells having elongated vesicular blunt-ended nuclei with inconspicuous nucleoli and brightly eosinophilic fibrillary cytoplasm without atypia. The cells were arranged in intersecting bundles and whorls encasing or surrounding numerous vascular channels. The smooth musculature of the vessel walls seemed to merge gradually and imperceptibly with the surrounding smooth muscle bundles. There was no cellular pleomorphism and mitotic figures were absent. Twelve of 16 cases showed compact arrangement of the smooth muscle occasionally mimicking leiomyoma but careful assessment revealed the characteristic thick-walled collapsed vascular channels (solid type) (Fig. 2a). Two lesions showed convolutes of thick-walled venous channels closely mimicking venous hemangioma (venous type) (Fig. 2b). Another two cases showed prominent dilated venous vessels within the smooth muscle proliferation corresponding to the cavernous type (Fig. 2c). A variable but generally prominent adipocytic component was appreciated in four cases (25 %). Mature macrovesicular adipocytes were scattered either singly or forming small aggregates and lobules within the lesions. The fatty

Table 1 Clinicopathological features of previously reported sinonasal angioleiomyomas including current series (n = 54)

No.	Author	Age/gender	Site, size cm	Symptoms, duration	Histological pattern	IHC positive markers	IHC negative markers	Associated mucosal changes	Follow up (months)
1	Maesaka et al. [9]	49 F	Vestibule	Facial pain	Vascular	ND	ND	NS	NR
2	Ram [10]	40 M	Right inferior turbinate, 2.5 cm	Nasal obstruction, few mo.	Solid (reported as fibromyoma)	ND	ND	NS	NA
3	Wolfowitz et al. [11]	42 F	Inferior turbinate, 1 cm	Recurrent epistaxis	Vascular leiomyoma	ND	ND	Ulceration	NR (30 mo.)
4	Schwartzman et al. [12]	57 M	Sinuses	Obstruction, headache	Vascular	ND	ND	NS	NR (36 mo.)
5	McCafferey et al. [13]	76 F	Inferior turbinate, 0.5 cm	Epistaxis	Vascular	ND	ND	NS	NA
6	Dawlatly et al. [14]	52 M	Right vestibule, 4 cm	Epistaxis, obstruction, 1 year	Solid with fat	ND	ND	None	NR (12 mo.)
7	Hanna et al. [15]	64 F	Inferior turbinate, 3 cm	Epistaxis, obstruction, pain, 3 wks	Solid, no fat	ND	ND	None	NR (12 mo.)
8	Sawada [16]	41 M	Right nasal cavity/ orifice	Mass, slowly growing for years	Solid with fat	ND	ND	Erythema	NR (12 mo.)
9	Ragbeer et al. [17]	49 F	Right nasal floor, 1.5 cm	Epistaxis, pain, suppuration	Solid with fat	Desmin, MSA, vimentin	S100	None	NR (12 mo.)
10	Harcourt et al. [18]	55 F	Right ethmoid sinus, 2 cm	Right epiphora, 10 years	Venous, no fat	ND	ND	ND	NR
11	Khan et al. [19]	71 F	Left inferior turbinate, 4 cm	Obstruction, 2 years	Solid with fat	NS	NS	NS	NR (12 mo.)
12	Gatalica et al. [20]	64 M	Right vestibule, 2 cm	Obstruction, >1 year	Solid with dominant fat	Muscle-specific actin, vimentin	HMB45, desmin	NS	NA
13	Ardekian et al. [21]	54 F	Left nasal vestibule and septum, 2 cm	Obstruction, pain and bloody discharge	Solid, no fat	NS	NS	Inflammation	NA
14	Nall et al. [22]	43 F	Superior turbinate	Epistaxis, obstruction, facial pain, months	Venous	αSMA	NS	No	NR (21 mo.)
15	Murono et al. [23]	69 F	Right inferior turbinate, 2 cm	Epistaxis, 1 year	Solid with fat	Vimentin, αSMA	NS	No	NA
16	Watanabe et al. [24]	66 M	Right nasal cavity, 2 cm	Mass, after chemotherapy for Hodgkin lymphoma	Solid with fat	αSMA, MSA, focally desmin	S100, HMB45	No	NR (24 mo.)
17	Watanabe et al. [24]	88 F	Vestibule, 2 cm	Mass	Solid with fat	αSMA, MSA, focally desmin	S100, HMB45	No	NR (6 mo.)
18	Marioni et al. [25]	70 F	Right vestibule, 1.5 cm	Obstruction and epistaxis	Solid-venous, no fat	αSMA, PR	ER	No	NR (3 mo.)
19	Tardio et al. [26]	45 M	Right nasal cavity, 1.5 cm	Epistaxis and obstruction, 2 mo.	Solid with fat	αSMA, desmin, MSA	HMB45	No	NR (1 mo.)

Table 1 continued

No.	Author	Age/gender	Site, size cm	Symptoms, duration	Histological pattern	IHC positive markers	IHC negative markers	Associated mucosal changes	Follow up (months)
20	Wang et al. [27]	70 M	Septum, 1.1 cm	Epistaxis	Solid, no fat	SMA	NS	NS	NR
21	Wang et al. [27]	66 F	Inferior turbinate, 0.3 cm	Asymptomatic mass	Venous, with fat	αSMA	NS	NS	NR
22	Bel Hag Salah et al. [28]	50 F	Right middle turbinate	Nasal obstruction + rhinorrhea, 5 mo.	Solid, no fat	αSMA, h-caldesmon	NS	NS	NA
23	Erkilic et al. [29]	52 M	Left nasal cavity, 3.5 cm	Snoring and obstruction, 20 years	Solid with fat	αSMA	S100, HMB45	NS	NA
24	Chen et al. [30]	88 M	Right inferior turbinate, 1.3 cm	Discharge, hearing impairment	Solid with fat	αSMA, desmin	ER, PR, CD34, EBV ISH	Inflammation	NR (12 mo.)
25	Meher et al. [31]	24 F	Right middle turbinate, 2 cm	Epistaxis, 2 mo.	Solid-venous, no fat	NS	NS	NS	NR
26	Campelo et al. [32]	44 F	Left turbinate	Recurrent epistaxis (4 years), obstruction (1 year), itching	Solid, no fat	NS	NS	NS	NR (10 mo.)
27	Vafiadis et al. [33]	68 M	Right nasal vestibule, 2 cm	Nasal obstruction, >6 years	Solid, no fat glands entrapped	NS	NS	No	NR (24 mo.)
28	Singh et al. [34]	31 M	Right nasal cavity septum, 3.2 cm	Nasal obstruction + intermittent epistaxis, 3 years	Solid, no fat	NS	NS	Surface ulceration	NR (18 mo.)
29	Michael et al. [35]	34 M	Left nasal cavity inferior turbinate	Nasal obstruction + intermittent epistaxis, 10 years	Venous	αSMA, desmin	NS	NS	NA
30	He et al. [36]	58 M	Right inferior turbinate, 2 cm	Recurrent epistaxis and obstruction, 10 years	Solid with fat	PR (20–30 %)	ER, HMB45, EBV	Erosion and inflammation	NR (12 mo.)
31	Navarro et al. [37]	62 F	Left septum, 4 cm	Epistaxis, obstruction, pain, mass, 6 years	Solid, no fat	NS	NS	NS	NA
32	Moreira et al. [38]	54 M	Left inferior meatus	Recurrent epistaxis, 20 years	Solid with fat	NS	HMB45	NS	NR
33	Purohit et al. [39]	45 M	Septum	Nasal obstruction, 3 years	Solid, no fat	NS	NS	No	NA
34	Yoon et al. [40]	64 M	Inferior turbinate, 0.8 cm	Mass	Solid	Actin+	NS	NS	NR
35	Yoon et al. [40]	65 M	Nasal mucosa, 1 cm	Mass	Solid	Actin+	NS	NS	NR
36	Yoon et al. [40]	37 M	Nasal septum, 1 cm	Epistaxis	Cavernous	Actin+	NS	NS	NR
37	Yoon et al. [40]	71 F	Nasal septum, 2 cm	Nasal obstruction	Cavernous	ND	NS	NS	NR

Table 1 continued

No.	Author	Age/gender	Site, size cm	Symptoms, duration	Histological pattern	IHC positive markers	IHC negative markers	Associated mucosal changes	Follow up (months)
38	Tseng et al. [41]	48 F	Right inferior turbinate, 1 cm	Recurrent mucous + bloody discharge, 3 mo.	Solid, no fat	αSMA, PR	ER, S100	No	NR (48 mo.)
39	Current study	73 M	Right lateral nasal wall, 1.4 cm	Intermittent nasal obstruction, 10 years.	Solid with fat (<10 %)	Desmin, αSMA, h-caldesmon	HMB45	None	NR (52 mo.)
40	Current study	82 M	Lower left turbinate, 0.8 cm	Recurrent epistaxis for years	Solid with fat (<10 %)	αSMA, h-caldesmon	HMB45, desmin	Erosion with inflammation	NR (43 mo.)
41	Current study	53 M	Left nasal orifice, 0.8 cm	Mass	Venous, no fat	αSMA, h-CD, desmin ±, CD56±	AR, HMB45, D2-40	Erosion with inflammation	NR (34 mo.)
42	Current study	76 F	Right nasal orifice, 0.6 cm	Mass	Solid with fat (40 %)	Desmin, αSMA, h-caldesmon	HMB45, ER, PR, CD56	None	NR (32 mo)
43	Current study	63 M	Right septum, 0.7 cm	Tumor, pyogenic granuloma?	Cavernous with fat (<2 %)	Desmin, αSMA, h-caldesmon	HMB45, CD56, D2-40	None	NR (31 mo.)
44	Current study	25 F	Ethmoidal cells, 0.2 cm	Incidental, recurrent sinusitis	Venous, No fat	Desmin, αSMA, h-caldesmon	HMB45	None	NR (15 mo.)
45	Current study	77 F	Nasal cavity, 0.7 cm	Mass	Solid with fat (<10 %)	αSMA, h-caldesmon, CD56	Desmin, HMB45	Erosion with inflammation	NR (211 mo.)
46	Current study	62 F	Nasal cavity, 1.5 cm	Mass	Solid, no fat	αSMA, desmin F +, h-caldesmon, CD56	HMB45	Ulcerated with inflammation	NR (80 mo.)
47	Current study	48 F	Concha, 1.2 cm	Mass	Solid, no fat	αSMA, desmin, h-caldesmon, CD56	HMB45	Ulcerated with inflammation	NR (161 mo.)
48	Current study	26 M	Right inferior nasal cavity floor, 2.5 cm	Painful mass, increasing in size, 3 mo	Solid, no fat	αSMA, MSA, desmin	S100 protein, pan-cytokeratin, HMB45	None	NR (108 mo.)
49	Current study	55 M	Right septum, 1.0 cm	Multiple polyps, nasal congestion, sneezing, difficulty breathing, pan-sinusitis and anterior septum mass, 1.5 mo.	Solid, no fat	MSA	S100 protein	None	NR (53 mo.)
50	Current study	77 F	Right anterior turbinate, 0.9 cm	Epistaxis with friable mass, 9 mo.	Solid, no fat	Desmin	HMB45	None	NR (46 mo.)
51	Current study	51 F	Left nasal polyp, 1.7 cm	Ear pain, cough, foreign-body sensation, nasal obstruction and epistaxis, 8 mo.	Cavernous, no fat	αSMA, desmin	S100 protein, CD34, pan-cytokeratin	Surface ulceration (excoriation)	NR (26 mo.)
52	Current study	36 M	Right lateral nasal wall (turbinate), 1.7 cm	Mass at the nasal vestibule, showing recent growth, 8 mo.	Solid, no fat	αSMA, SMMHC	Desmin, S100 protein	None	NR (20 mo.)

Table 1 continued

No.	Author	Age/gender	Site, size cm	Symptoms, duration	Histological pattern	IHC positive markers	IHC negative markers	Associated mucosal changes	Follow up (months)
53	Current study	65 M	Right anterior nasal septum, 1.0 cm	Nasal congestion, with a past history of rhinoplasty and difficulty breathing and epistaxis, 20 mo.	Solid, no fat	MSA	none	Squamous metaplasia	NR (18 mo.)
54	Current study	66 M	Right inferior turbinate, 1.2 cm	Right nasal obstruction with epistaxis, 24 mo.	Solid, no fat	MSA, desmin	S100 protein, EBER	Squamous metaplasia	NR (9 mo.)

F female, *M* male, *MO* month, *MSA* muscle-specific actin, *NA* not available, *NR* no recurrence, *PR* progesterone receptor, *SMA* smooth muscle actin, *SMMHC* smooth muscle myosin heavy chain

component ranged from a few cells to 20 % of the lesion (Fig. 3d–h). There were no cytoplasmic vacuoles within the smooth muscle cells or other features suggestive of gradual transition from myogenic to fatty cells. The Elastica stain confirmed the venous nature of the vessels (Fig. 3a).

By immunohistochemistry, all cases tested showed strong expression of alpha smooth muscle actin (12/12), h-caldesmon (9/9), muscle-specific actin (4/4) and variable expression of desmin (11/14; diffuse in nine cases, focal in two and negative in three cases). Six cases were tested for CD56: three showed a diffuse reaction, one was focally positive, and two were negative. No correlation between CD56, desmin, and histological pattern was observed for the cases stained for both markers. Two cases stained with D2-40 showed no lymphatic component. The proliferation fraction (Ki-67) was <2 %. None of the 11 cases tested were reactive for HMB45. Selected immunohistochemistry findings are illustrated in Fig. 3b–d.

Discussion

Head and neck angioleiomyoma (ALM) are rare, comprising 8.5 and 13 % of all ALMs in two larger series [7, 8]. ALMs originating from sinonasal mucosa-covered sites are even rarer. They represented 9.5–12.5 % of head and neck and 1 % of all ALMs, respectively [7, 8, 27]. Only a single case of ALM was identified among 331 consecutive benign sinonasal masses (0.3 %) [42]. Since the first description by Maesaka et al. [9], no more than 38 well documented cases have been reported in the English literature, mainly as single case reports [9–41]. Although a female predilection has been suggested [1], review of previously reported cases combined with this clinical series (total: 54; Table 1) showed that both genders are affected equally (28 males and 26 females). Age range of reported cases was 24–88 years (mean 57 years). Mean age is 56 and 55 years for women and men, respectively. The turbinates are affected most frequently, followed by other subregions of the nasal cavity including nasal orifices, septum and lateral nasal wall. The paranasal sinuses were affected in only three cases. Most sinonasal ALMs present as sessile or polypoid well circumscribed but non-encapsulated masses. Their size ranged from 0.2 to 4 cm (mean 1.7 cm). Seven of 45 cases (15 %) with detailed information measured >2 cm. Pain, a characteristic symptom of cutaneous vascular leiomyoma [1, 6, 7] was reported in 5/54 patients with sinonasal tract ALM. Another three patients reported facial pain/headache, among other symptoms.

An infrequent finding in ALMs in general is the observation of a variable clustered or intermingled component of mature adipocytes, which resulted in the use of the

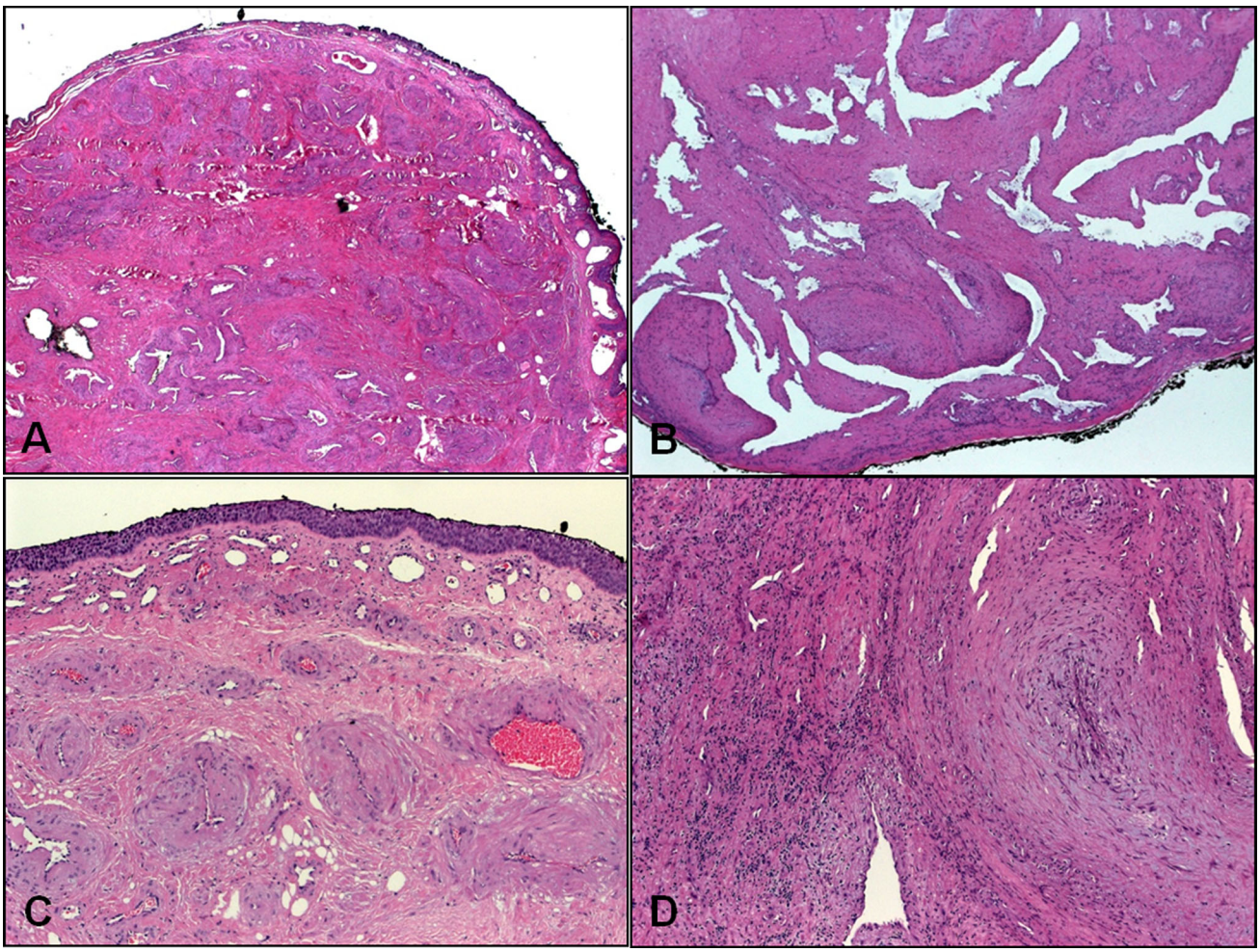
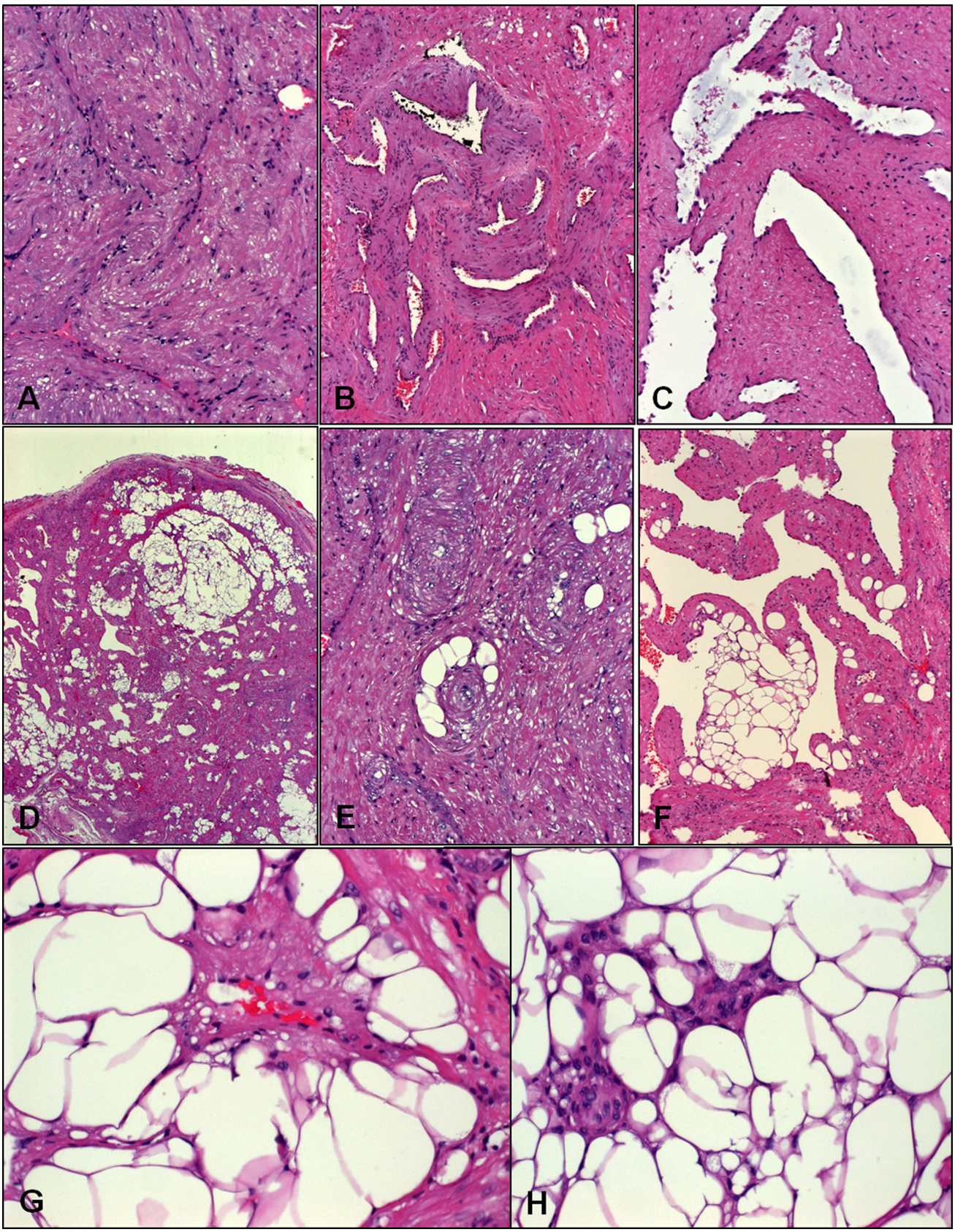


Fig. 1 Low-power findings in sinonasal angioleiomyomas. **a** Compact submucosal growth of haphazardly arranged thick-walled venous vessel with intervening fibromuscular stroma. **b** This example showed ectatic (cavernous) venous channels with their muscular walls blending with background musculature. **c** Surface epithelium showed squamoid metaplasia. **d** Interstitial inflammation with fibromyxoid vascular obliteration in ulcerated lesions may mask the underlying tumor

alternative term *angiomyolipoma* for some of previously reported cases [14, 24, 26, 29]. Among ALMs from all sites, a fatty component was observed in 2.8 % of cases [7]. However, review of reported cases and this clinical series (Table 1) showed a higher frequency of fatty component in sinonasal submucosal ALMs compared to their cutaneous counterparts (35 vs. 2.8 %, respectively). Comparing the subcohorts with (n = 19) and without (n = 35) adipocytic differentiation, ALM with fat tends to affect males more frequently than those without fat (63 vs. 46 % males) and to occur at a higher age (65 vs. 52 years for those with and without fat, respectively). Mean size was similar in both subgroups (18 and 16 mm, respectively).

Given that the presence or absence of a fatty component was not mentioned in several of the previously reported cases, it is possible that the frequency of adipocytic differentiation is even higher. None of the cases with available immunohistochemical findings stained for the melanocytic marker HMB45. The histological features were uniformly those of cutaneous-type ALM with or without adipocytic differentiation [43]. Based on these reported cases, sinonasal ALMs with adipocytic differentiation are histologically identical to their cutaneous/soft tissue counterparts and are distinctly different from renal-type angiomyolipoma. The latter can be associated with tuberous sclerosis complex, shows characteristic granular myogenic cells with frequent perivascular aggregates of epithelioid cells, convoluted thick-walled dysplastic vessels and cells with intermediate or transitional features between mature fatty cells and myogenic cells with cytoplasmic vacuoles indicating continuous differentiation. By immunohistochemistry, angiomyolipoma of renal type characteristically co-expresses myogenic and melanocytic markers (mainly HMB45). On the other hand, sinonasal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◀**Fig. 2** Histological spectrum of sinonasal angioleiomyomas. **a** Solid type with collapsed vascular channels amid smooth muscle bundles. **b** Lobule-like convolutes of veins surrounded by smooth muscle stroma. **c** This lesion showed ectatic vascular channels enclosed within smooth muscle bundles without discernible vascular walls. **d** Overview of a fat-rich lesion. **e** This solid lesion contained scattered adipocytes forming ring-like aggregates surrounding vessels. **f** Cavernous lesion with fatty lobules within vascular walls. **g** Perivascular "ring-adipocytes" from another case. **h** This fat-rich lesion showed size variation of adipocytes and can be mistaken for angiomyolipoma or adipocytic neoplasm

ALM shows a mature smooth muscle phenotype (desmin+/α-SMA+/h-caldesmon+/HMB45−). On critical review of the literature, it is evident that many of the reported sinonasal angiomyolipomas were actually "ALM with adipocytic differentiation" [14, 24, 26, 29], but a few cases of genuine renal-type angiomyolipomas and PEComas have been documented in the sinonasal tract as well [44, 45]. Accordingly, ambiguous and misleading terms such as angiomyolipoma and angiolipoleiomyoma should be abandoned. Instead, the term "angioleiomyoma with adipocytic differentiation" should be used for sinonasal ALMs with fatty component, analogous to their cutaneous and soft tissue counterparts [46]. Some ALMs may show focal perivascular concentric myoid cells closely mimicking myopericytoma [43]. On the other hand, the presence of ALM-like myopericytoma is well appreciated [47]. While most myopericytomas are composed of alpha smooth muscle actin+, h-caldesmon+, desmin-negative perivascular myoid cells, 15–20 % of ALMs are desmin-negative as observed in previous studies [43] and in our current series. These observations suggest the presence of lesions with overlapping features of ALM and myopericytoma, at least in a subset of cases [43, 47]. In our current series, none of the cases showed myopericytoma-like features. Albeit rare, Epstein-Barr Virus (EBV)-associated smooth

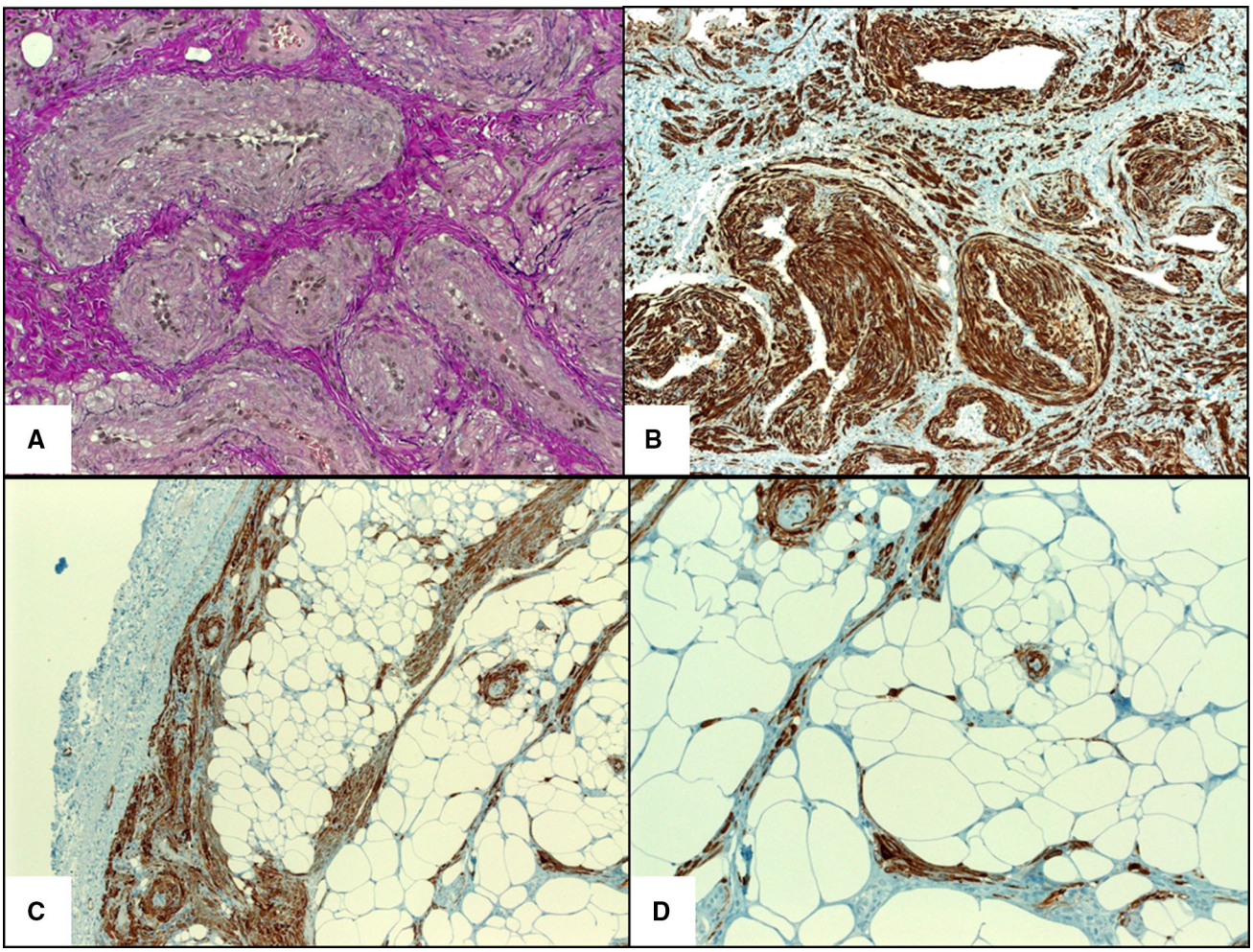

Fig. 3 **a** Elastic van Gieson stain highlighting the venous channels. **b** Strong desmin expression highlighting intervascular smooth muscle bundles. **c** H-caldesmon showed strong reactivity in muscle cells (note peripheral circumscription). **d** Higher magnification of h-caldesmon showed scattered isolated smooth muscle cells amid the fatty component

muscle tumors may on occasion closely mimic ALM. These rare lesions are characteristically multifocal involving different organs and affect patients with acquired or congenital immunodeficiency. Definitionally, they harbor EBV, detectable by in situ hybridization methods for EBER [48].

Review of the previous cases showed that some lesions likely represented other entities (excluded in the current review). In particular, sinonasal glomangiopericytoma/hemangiopericytoma was more commonly confused with ALM, given that both lesions strongly express smooth muscle actin. Sinonasal glomangiopericytoma, however, is a cellular neoplasm composed of monomorphic short spindled or ovoid to glomoid cells with distinctive cytoplasm and characteristic pericytomatous vascular pattern. They usually show a characteristic peritheliomatous hyalinization, lacking in ALM. Further, they lack the cytoplasmic eosinophilia and elongated blunt-ended nuclei of mature smooth muscle cells of ALMs and they are negative for h-caldesmon and desmin [3]. Two recent studies showed uniform nuclear expression of β-catenin in sinonasal glomangiopericytoma as a consequence of β-catenin mutations [49, 50].

In summary, we reported 16 new cases and reviewed 38 previously reported cases of submucosal sinonasal angioleiomyomas emphasizing their benign nature, frequent fatty component, similarity to cutaneous angioleiomyoma and distinctness from renal-type angiomyolipoma and equal gender distribution. Awareness of their histological spectrum is necessary to distinguish them from other potentially recurring or locally aggressive neoplasms.

Conflict of interest None.

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