CASE REPORT

Leiomyoma: A rare tumor in the head and neck and oral cavity: Report of 3 cases with review

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ABSTRACT

Leiomyomas are benign tumors arising from smooth muscle, most commonly seen in uterine myometrium, gastrointestinal tract, skin and lower extremities of middle-aged women. Leiomyomas are uncommon in the oral cavity with reported incidence of 0.065%, which accounts for 0.42% of all soft-tissue neoplasms in the oral cavity. Leiomyomas of head and neck region account for less than 1% of all leiomyomas. The most common site of leiomyoma in the head and neck region is the lips (27.46%) followed by tongue (18.30%), cheeks and palate (15.49%), gingiva (8.45%) and mandible (5.63%). The purpose of this article is to present three cases of leiomyoma comprising of an intraoral vascular leiomyoma and two solid leiomyomas in the head and neck region. The clinical features, etiology, differential diagnosis and treatment of leiomyoma are discussed with review of the literature.

Key words: Angioleiomyoma, benign, ear auricle, intraoral, leiomyoma, smooth muscle, solid variant

INTRODUCTION

Leiomyoma is a benign soft-tissue neoplasm arising from smooth muscle. It was first described by Virchow in 1854 and first reported by Blanc in 1884. The hereditary form, which causes multiple leiomyomas was originally noted by Kloepfer *et al.*, in 1958.[1] Since head and neck region lacks smooth muscle, occurrence of leiomyoma in this area is rare. Leiomyomas are most commonly found in uterine myometrium (95%) followed by skin (3%), gastrointestinal tract (1.5%) and lesser than 1% are seen in the head and neck region. Incidence of leiomyoma in the oral cavity is 0.065%.[2,3] The tumor is commonly seen in 4th and 5th decade with slight predilection for females and generally manifests as a slow-growing, asymptomatic lesion.

Clinically, connective tissue tumors e.g. fibromas, lipomas may present similar findings to leiomyoma; hence a differential diagnosis must be established. Histological findings play a key role in the final diagnosis of leiomyoma. The differential diagnosis, moreover, must also include the malignant form of leiomyoma, i.e. leiomyosarcoma. At present, surgical resection is the main treatment for leiomyoma and recurrences are extremely rare.

This article will elaborate on three cases of leiomyoma arising in the head and neck region. Two cases were found associated with ear auricle while one was found intraorally in relation to the hard palate.

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CASE REPORTS

Case 1

A 25-year-old female patient reported to out-patient department with a chief complaint of swelling in the right lower portion of the ear since 6 months. The swelling was painless, asymptomatic, slow growing and esthetically unacceptable for the patient.

Clinical findings

The swelling was solitary, ovoid in shape, involving the right ear lobe, measuring about 1.5 cm × 1 cm, covered by normal appearing skin, which was stretched and mobile. It was firm in consistency having well-defined margins. Trans-illumination test was negative [Figure 1].

Surgical resection was planned with an excisional biopsy. An elliptical incision was placed and the lesion was excised under local anesthesia [Figure 2]. The post-operative period was

uneventful with 1 year follow-up and there was no sign of recurrence.

Histopathological findings

The tissue specimen showed a non-encapsulated and well-circumscribed lesion. The cells were arranged in whorls and fascicles [Figure 3]. The cells were spindle shaped with eosinophilic cytoplasm and centrally located basophilic, cigar shaped nucleus. Prominent vascular spaces were seen. There was no evidence of nuclear atypia between the smooth muscle bands; or necrosis or hemorrhage. Thus, the histological diagnosis of solid variant of leiomyoma was made.

Case 2

A 24-year-old female patient reported to out-patient department with a chief complaint of growth on the right pinna and ear lobule since 3 months. Two separate growths were seen associated with pinna and lobule of the right ear respectively; both of which were painless and had a gradual onset.

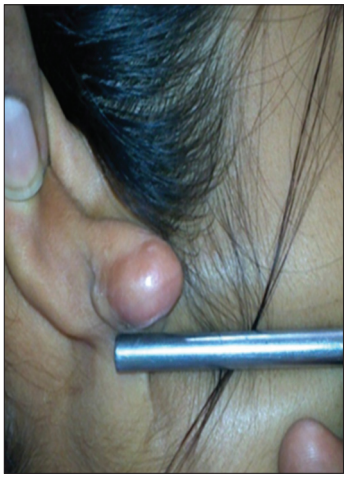

Figure 1: Clinical picture showing right ear lobe swelling

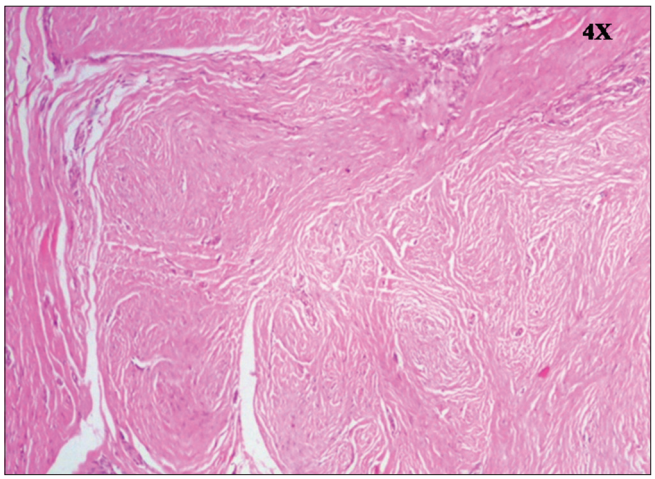

Figure 3: Photomicrograph showing a well circumscribed lesion having spindle shaped cells arranged in whorls and fascicles with eosinophilic cytoplasm (H&E stain, ×40)

Clinical findings

Clinical examination showed ovoid swellings measuring 2 cm × 1 cm and 1 cm × 1 cm respectively with well-defined margins [Figure 4]. Overlying skin appeared normal. Swellings were firm in consistency, non-tender and did not elicit fluctuation or translucency. The lesions appeared to be fixed to the underlying cartilage.

Medical history and family history was not contributory and thus syndromic or hereditary association was ruled out.

Surgical resection of both swellings was performed under local anesthesia. Resected specimens were sent for histopathological examination. Regular follow-up for a period of 3 years did not show any signs of recurrence.

Histopathological findings

The histopathological examination of the hematoxylin and eosin stained slides showed moderately cellular lesional tissue comprising of spindle cells with indistinct cytoplasmic membrane and nuclei having blunt ends [Figure 5]. Both longitudinal areas running in fascicles (hypercellular) and cross-sectional areas (hypocellular) were evident with numerous endothelial lined blood vessels [Figure 6]. There

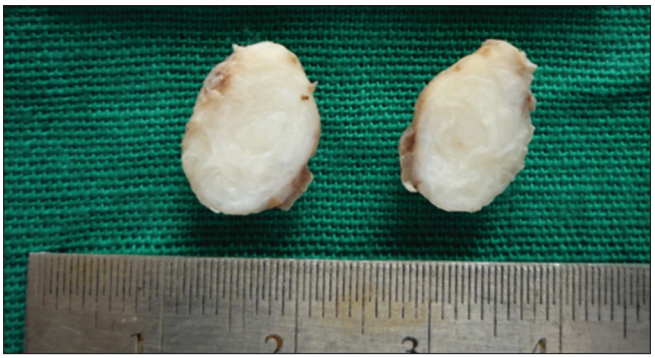

Figure 2: The cut surface of the excised mass

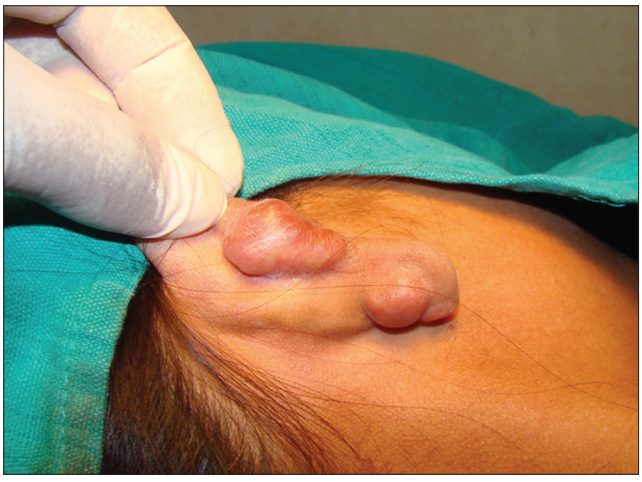

Figure 4: Clinical picture showing two well defined growths on right auricle

were no features of cellular atypia or necrosis. Thus, a histological diagnosis of solid variant of leiomyoma was given.

Case 3

A 48-year-old male patient was referred to our department with a chief complaint of swelling in relation to right upper molar teeth since 2 months.

Clinical examination

On examination, a solitary swelling was noted on the right side of the posterior palate associated with right upper second premolar and first molar measuring 1 cm × 1.5 cm. Overlying mucosa was pale pink in color and appeared normal. The swelling was a sessile, firm, dome shaped mass fixed to the underlying tissues [Figure 7].

Aspiration biopsy was performed, which ruled out a vascular lesion. Radiographic picture showed no changes associated with underlying bone. The lesion was provisionally diagnosed as a minor salivary gland tumour. Excisional biopsy was performed and excised mass was sent for histopathologic examination [Figure 8].

Histopathological findings

The lesional tissue present in the deeper stroma was well-circumscribed and non-encapsulated and the cells were arranged in a streaming pattern around the blood vessels [Figure 9]. They formed whorls, which were separated by collagen bands. Numerous vascular spaces were seen surrounded by proliferating spindle cells [Figure 10]. The lesional cells were spindle in shape with blunt ended nucleus and had a moderate amount of cytoplasm with indistinct outline. There was no evidence of mitosis or necrosis. Thus, a diagnosis of angioleiomyoma was made.

Note: Special stains were done using Mallory's Phosphotungstic Acid-Hematoxylin for all the above three cases and the cells exhibited dark blue intracytoplasmic fibrils [Figure 11]. Masson trichrome staining exhibited smooth muscle cells (pink) in longitudinal and transverse sections with intervening collagen fibers (blue) [Figures 12-14].

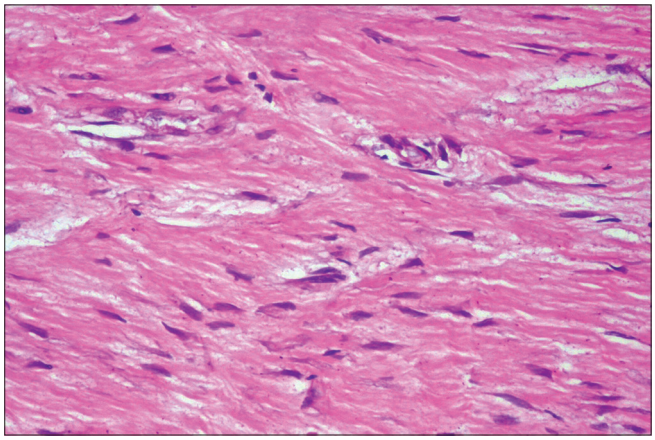

Figure 5: Photomicrograph showing spindle cells with indistinct cytoplasm and blunt ended nucleus (H&E stain, ×400)

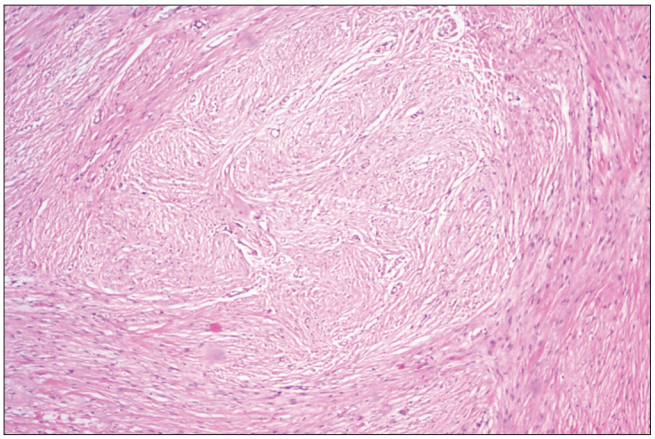

Figure 6: Photomicrograph showing spindle shaped cells arranged in fascicles with endothelial lined blood vessels (H&Estain, ×40)

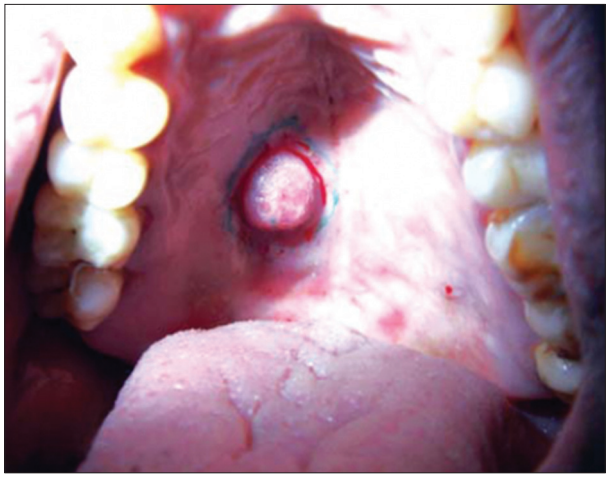

Figure 7: Clinical picture showing a palatal swelling

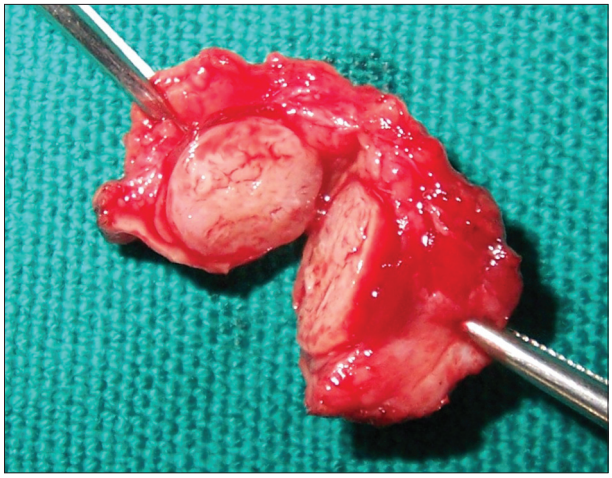

Figure 8: The cut surface of the lesion

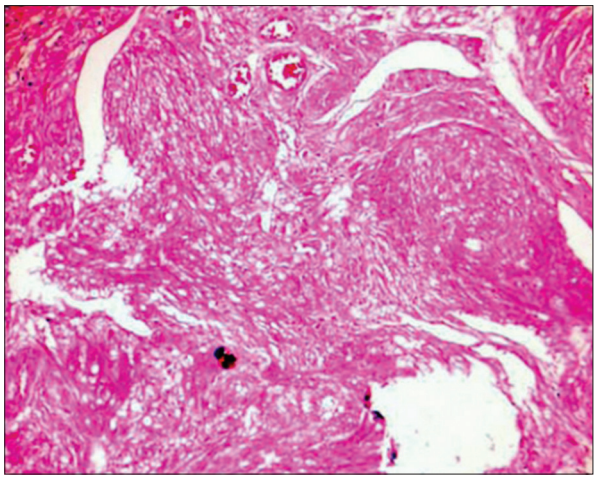

Figure 9: Photomicrograph showing plump or spindle shaped cells in a streaming pattern (H&E stain, ×200)

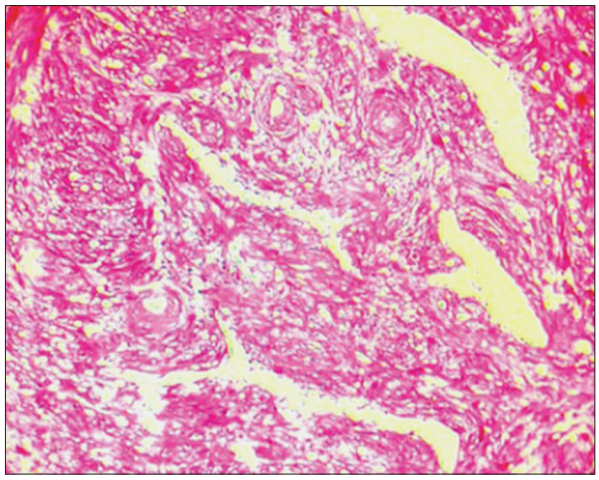

Figure 10: Photomicrograph showing arteries appearing as consumed by the spindle cells (H&E stain, ×200)

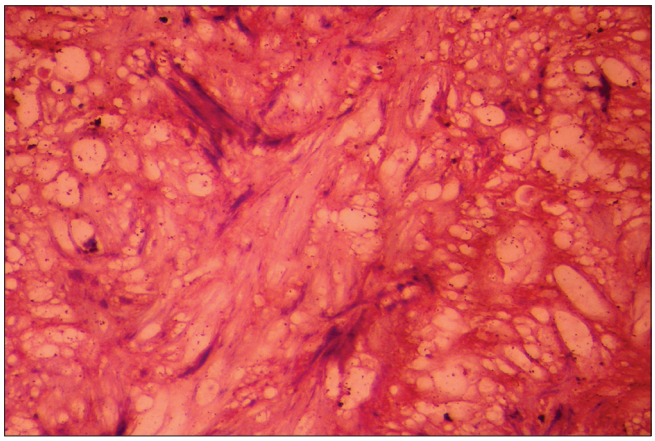

Figure 11: Photomicrograph showing the cells with dark blue staining intracytoplasmic fibrils (Mallory's phosphotungstic acid hematoxylin stain, ×400)

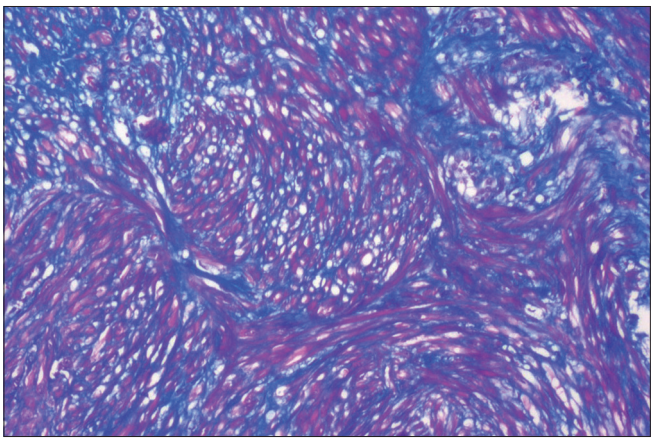

Figure 12: Photomicrograph showing smooth muscle fibers arranged in fascicles (pink) admixed with collagen fibers (blue) (Masson trichrome stain, ×100)

DISCUSSION

Leiomyoma is defined by the World Health Organization as a"circumscribed benign, often cutaneous tumor composed of intersecting bundles of mature smooth muscle cells."

Based on the histopathologic findings, the World Health Organization classified leiomyoma into three groups:[4] Solid leiomyoma, vascular leiomyoma (angioleiomyoma) and epithelioid leiomyoma (leiomyoblastoma). Both cases of leiomyomas involving the auricle described here are solid variants of leiomyoma.

Similarly oral leiomyomas are also classified as solid pattern leiomyomas, vascular leiomyomas and epitheloid leiomyomas.[4] The case of intraoral leiomyoma discussed here is a vascular leiomyoma also known as angioleiomyoma or angiomyoma.

Morimoto[5] in 1973 classified angioleiomyoma into three histologic subtypes: (1) Capillary or solid: The most common type, which shows closely compacted smooth muscle and many small, slit-like vascular channels. (2) Venous: Vascular channels with thick, easily identifiable muscular walls. (3) Cavernous: The vascular channels are dilated with less smooth muscle. Although capillary angioleiomyoma is the most common of the three subtypes, venous subtype is more commonly found in the oral cavity.[6]

Leiomyoma can appear in any location where smooth muscle is present. This lesion occurs most often in female genital tract. According to Enzinger and Weiss (1995), who analyzed a total of 7748 leiomyomas, 95% occurred in the female genitalia (uterus), 3% in the skin and the remainder in various other sites.[7]

In general, non-cutaneous head and neck leiomyomas are rare lesions. Only two leiomyomas were found by Fu and Perzin in a series review of 256 non-epithelial neoplasms of the sinonasal tract and nasopharynx.[8] Barnes reviewed 257 leiomyomas of head and neck and he found that 92 of

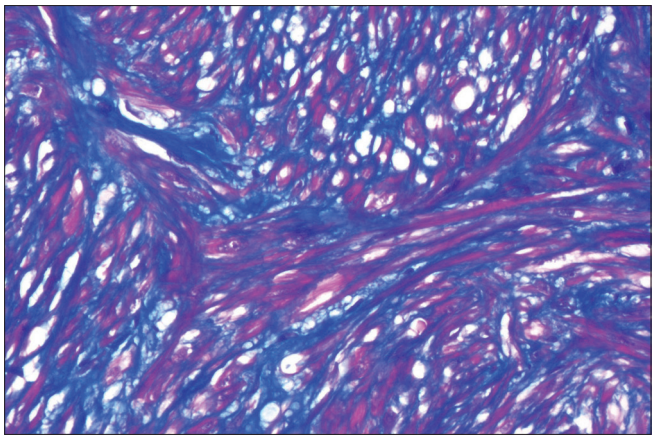

Figure 13: Photomicrograph showing smooth muscle cells (pink) in longitudinal and transverse sections with intervening collagen fibers (blue) (Masson trichrome stain, ×200)

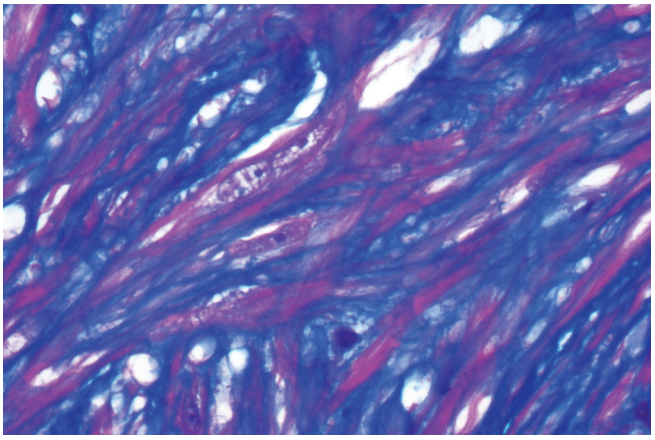

Figure 14: Photomicrograph showing smooth muscle cell (pink) with cigar shaped nuclei and a prominent nucleoli admixed with fibrous connective tissue (blue) (Masson trichrome stain, ×400)

them were associated with cervical esophagus, 58 with skin of head and neck, 52 with oral cavity, 22 with larynx, 12 with orbit, 6 with the nasal cavity and remaining were associated with trachea, salivary glands, paranasal sinuses, thyroid gland and the intraosseous ones associated with maxilla and mandible.[9] Of these 257 cases, 71% were solid variants, 27% were angioleiomyomas and 1.2% were epithelioid leiomyomas. Among the 58 leiomyomas associated with skin of head and neck, 22 were associated with ear or cheek followed by neck, forehead and scalp.[9]

Of the 52 leiomyomas found in the oral cavity, the most commonly involved site was lip (14) followed by tongue (13), palate (11), buccal mucosa (8), gingiva (4), buccal sulcus (1) and uvula (1).[9]

In head and neck, angioleiomyoma is found more frequently in the nasal cavity, paranasal sinuses and oral cavity. In a study of 562 cases of angioleiomyomas, Hachisuga *et al*., (1984) observed that 68% of the angiomyomas occurred in lower extremities, 22% in upper extremities and only 9% occurred in the head and neck.[10] Barnes reviewed 69 angioleiomyomas of head and neck and reported that 31 cases were found in the area of the oral cavity, most often in the lips (13), soft palate (8), tongue (5), buccal mucosa (3), gingiva (1) and uvula (1).[9]

The highest prevalence of head and neck leiomyomas is observed in 4th and 5th decades of life. The peak incidence is 40-49 years of age.[11] In our experience, the two cases of auricular leiomyoma were observed in a younger age group.

Higher incidence of this tumor (3.75:1=F:M) in females could be attributed to hormonal variations.[12] In recent years, sex steroid receptors (progesterone-receptor positive and oestrogen-receptor negative on immunohistochemical analysis) have been identified in leiomyomas, which suggest that the growth of these tumors could be hormone dependent.

Three hypotheses which explain the origin of smooth muscle tumors:[12]

1. From aberrant undifferentiated mesenchyme
2. From smooth muscle elements in the tunica media of blood vessels
3. Or from both sources.

Most lesions of leiomyomas in the head and neck region are asymptomatic, although occasional tumors can be painful. All our three cases presented as solitary, asymptomatic and slow growing lesions. Sinonasal leiomyomas typically manifest as polypoid nodular masses and may result in epistaxis, nasal obstruction, headache or occasionally facial pain.[8]

The clinical differential diagnosis relevant to extraoral leiomyoma include fibroma, lipoma, neurofibroma, dermatofibroma, lymphangioma, hemangioma and soft-tissue cysts such as dermoid cyst.[13]

Benign mesenchymal tumors such as fibroma, myofibroma, neurofibroma, schwannoma, lipoma; benign lesions of salivary gland origin, such as deep seated mucocele and pleomorphic adenoma should also be considered in differential diagnosis of intraoral leiomyoma. Similarly, vascular lesions such as hemangioma, pyogenic granuloma, lymphangioma; or malignant ones such as leiomyosarcoma are some of the oral tumors, which can present as leiomyoma. Therefore, these should be excluded before making the final diagnosis.[13]

Histolopathologically, differential diagnosis includes benign spindle cell tumors such as schwannoma, neurofibroma, fibrous histiocytoma, nodular fascitis and malignant ones such as leiomyosarcoma.

Schwannoma is an encapsulated lesion and has more cellularity comprising of schwann cells. It exhibits Antoni A areas with Verocay bodies and Antoni B areas that have microcystic

spaces and myxoid areas. The nucleus of these spindle cells is pointed and twisted. Immunohistochemistry (IHC) shows uniform strong positivity for S-100.

Neurofibroma, on the other hand, is unencapsulated tumor of neural (schwann) cells and fibrous tissue (fibroblasts) wherein the spindle shaped neural cells have pointed/tapered ends and are wavy in nature. Fibrous component exhibits a typical "shredded carrot" appearance. Mast cells and neurites are seen. The stroma is fibromyxoid in nature. IHC shows focal positivity for S-100.

Fibroma is a benign fibrous tumor, which is composed of fibroblasts. Collagen is arranged in varying patterns. Owing to secondary effects of proliferating fibroblasts, the overlying epithelium is atrophic and stretched.

Fibromatosis is a fibroblastic lesion with varying amounts of collagen. It is highly cellular tumor that is infiltrative and aggressive in nature, but cytologically bland. In spite of being cytologically bland most of these tumors show the presence of mitosis.

Leiomyomas must be carefully distinguished from their malignant counterpart - leiomyosarcoma, as both can demonstrate a similar clinical picture. Leiomyosarcoma although presents with similar histologic features as leiomyoma; atypia, cellularity, pleomorphism and necrosis may provide some indication of malignant disease and mitotic activity represents the most reliable criterion of malignant behavior.[13] Kumar and Cotran[14] stated that histological features indicative of smooth muscle malignancy include more than 10 mitoses/10 high-power fields (HPF) with or without cellular atypia and 5-10 mitoses/10 HPF with atypia. Tumors having 1-4 mitoses are best considered as potentially malignant especially if they are large and have areas of necrosis and significant nuclear atypia.

There is a prevailing view that leiomyomas of deep soft tissues are rare or non-existent, but there is limited data on this subject in the form of large clinical studies with long-term follow-up. After reviewing 36 consultation cases, Billings *et al*.,[15] concluded that clinically benign smooth muscle tumors of deep soft-tissues are rare but can be identified using the stringent histologic criteria. They comprise two distinct subtypes: Leiomyomas of somatic soft-tissue and retroperitoneal-abdominal leiomyomas.[15]

Intraosseous leiomyomas are rare. Some of the possibilities with respect to origin are smooth muscle of vascular walls and heterotopic embryonal tissue. Owing to its intraosseous rarity, leiomyoma is generally not considered in the radiographic differential diagnosis. More likely diagnostic considerations include central giant cell lesion, ameloblastoma, myxoma, traumatic bone cyst, hemangioma, neurofibroma and sarcoma.

Radiographically, angioleiomyomas manifest as unilocular or multilocular radiolucent lesions with either an ill-defined or a well-defined sclerotic border. Cortical expansion of the alveolar plates and root resorption may be evident.[16]

The treatment of choice is local resection, including an adequate safety margin of normal-appearing tissue. Despite the vascular origin of these lesions, excessive bleeding after excision is rare. Recurrence following complete resection is notably rare with leiomyoma.

CONCLUSION

Leiomyoma is a benign tumor of rare incidence in the head and neck regions and with a good prognosis. The diagnosis is mainly based on histological features. The treatment of choice is surgical excision with adequate safety margins. We have compiled three cases of leiomyoma to add to the existing literature.

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