Bucco-Parapharyngeal Occurrence of Pleomorphic Spindle Cell Lipoma-A Unique Entity: Case Report

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Lipomas are common benign neoplasms of oral region which occur scarcely in the parapharyngeal space (PPS). The diversity observed in its histologic presentation classified it into many subtypes among which Spindle Cell Lipoma (SCL) is a rare entity. SCL is an uncommon benign tumor of adipose tissue usually located superficially in the neck, back and shoulder region but its presentation in oral and pharyngeal spaces is very rare. Exhaustive literature search for oral and pharyngeal space pleomorphic SCL till date revealed 45 cases of oral SCL with a mean age distribution of 55 years. Only one reported case of parapharyngeal occurrence of pleomorphic SCL was found. A rare case of pleomorphic variant of SCL in PPS extending into the buccal space is being presented. This case could be the first to report the multiple presentation and the youngest age of occurrence of SCL in PPS.

***Keywords**: Pleomorphic spindle cell lipoma; Floret cells; Parapharyngeal space; Buccal space.*

INTRODUCTION

Lipomas, the benign neoplasms of adipose tissue are the most common soft tissue tumors, usually occurring in upper back, shoulder, abdomen, head, neck, and proximal extremities.[1-3] In the oral region they are uncommon and comprise 4%–5% of all tumors.[1] Histologically, lipomas are classified into various subtypes, including fibrolipoma, angiolipoma, myxoid lipoma, spindle cell lipoma (SCL), pleomorphic lipoma (PL), myolipoma, and chondroid lipoma.[1] In oral cavity, classic lipomas are the most common subtype, followed by SCLs, fibrolipomas, chondroid lipomas, and angiolipomas. SCL is an uncommon variant, originally described in 1975 by Enzinger and Harvey.[4] SCL usually presents in the subcutaneous tissue of the posterior neck, shoulder, and back, but rarely it is been reported to occur in other sites, like the oral cavity, spermatic cord, hypopharynx, parotid gland, vulva, limbs, and arms and accounts for approximately 1.5% of all lipoma cases.[5] Oral SCL is considerably rare with an average incidence of about 7%, ranging between 2.17% and 9.8% of all oral lipomas.[1] Parapharyngeal space (PPS) tumors are very rare, accounting for about 0.5% of head and neck tumors. Although 13% of the lipomas are seen in the head and neck region, only 1% to 2% are seen in the PPS. Almost, 50% of the PPS neoplasms are of salivary gland origin followed by 30% originating from the neurogenic tissue and lipomas being very rare in this space. Histopathologically SCL is characterized by a mixture of mature adipose tissue, bland spindle cells and ropy collagen in focal myxoid stroma. The pleomorphic variant has a characteristic presence of "floret type" giant cells.[1,2,4,5] Here we describe a rare case of pleomorphic variant of SCL with a unique location i.e. left buccal and parapharyngeal spaces in a 15 year old boy. Differential diagnosis of pleomorphic variant of SCL is emphasised based on its histological and immunohistochemical characteristics.

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Case Report

A 15 year old boy reported to the outpatient department with a slow growing painless swelling on left cheek region discerned for the last 2 months, which was initially small, asymptomatic and increased to present size by time. There was no family or medical history relatable to this condition and neither any history of trauma. Patient was well built and nourished with healthy mental orientation. On clinical examination a single swelling was noted in left cheek region appearing ovoid in shape with an approximate size of 2.5×3 cms with normal appearing overlying skin (Figure 1). On palpation the swelling felt soft in consistency, non-tender, compressible and fluctuant with feebly appreciable pulsation. Mouth opening was reduced which the patient claimed to experience after the swelling. Intraorally **it** obliterated the left posterior vestibular space. The buccal mucosa and gingiva along the side of swelling appeared normal. Radiographic findings by magnetic resonance imaging (MRI) revealed two well-encapsulated hyperintense masses, one in buccal space(Figure 2A) and other in PPS (Figure 2B) with no evidence of any mandibular bone involvement. Absence of feeder vessels by the computed tomography angiography(CT angiography) ruled out the probability of any lesion of vascular origin. The lesion was surgically excised under general anaesthesia and sent for histopathological examination with a provisional diagnosis of lymphangiohemangioma (Figure 3A). Taking into account the clinical, radiological features as well as location, age and presentation of lesion differential diagnosis of salivary gland, neurogenic or any adipose tissue tumors were listed. Gross examination of biopsy specimen recorded two separate round to ovoid masses, (buccal and parapharyngeal) joined by a single capsule with an approximate size of 3×3×3 cms and 5×4×3 cms respectively. The tissue was soft in consistency with the cut surface appearing creamish white and smooth(Figure 3B). After routine tissue processing, the histopathological examination of lesion from buccal space revealed an encapsulated mass containing lobules of mature adipocytes (Figure 4A) along with areas of myxoid changes (Figure 4B) and few foci of spindle cells, floret giant cells (Figure 4C) and ropy collagen fibers (Figure 4D). Undifferentiated spindle cells in sheets with floret giant cells & few adipocytes were seen in sections from the parapharyngeal lesion. Immunohistochemical analysis (IHC) was done using antibodies of S100, CD34, vimentin and desmin. On Immunohistochemical (IHC) analysis spindle cells were strongly immunoreactive for CD34 (Figure 5A) but negative for S100 (Figure 5B). Vimentin was positive for spindle cells (Figure 5C) whereas desmin was negative (Figure 5D) confirming its origin from mesenchymal cells to modified fibroblasts producing ropy collagen rather than a lipocyte. Adipocytic component was positive for S-100 protein while negative for CD34 and desmin (The IHC analysis results have been tabulated in Table 1). The histopathological and IHC analysis confirmed the diagnosis of pleomorphic variant of spindle cell lipoma. The present case has been under regular follow up for the past 14 months, shown complete healing of the surgical site and no signs of recurrence.

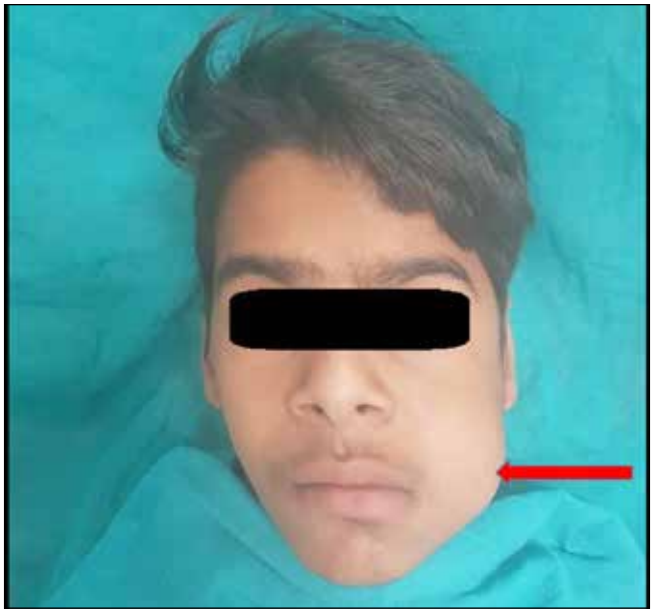

Figure 1: 1a) Clinical picture of the boy shows the extraoral swelling present on left side of face (arrow)

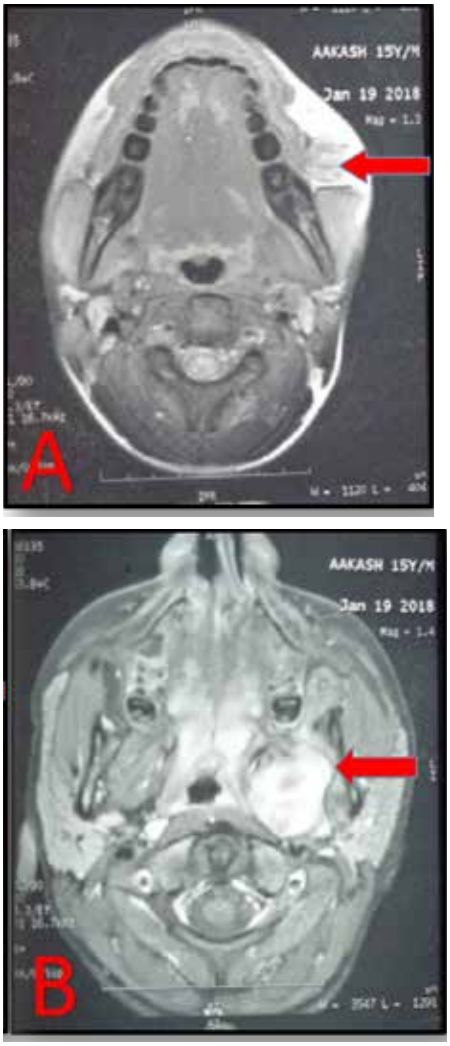

Figure 2: 2a) Magnetic resonance imaging (MRI) reveals a hyperintense mass in both buccal (arrow) and 2b) mass in parapharyngeal space (arrow).

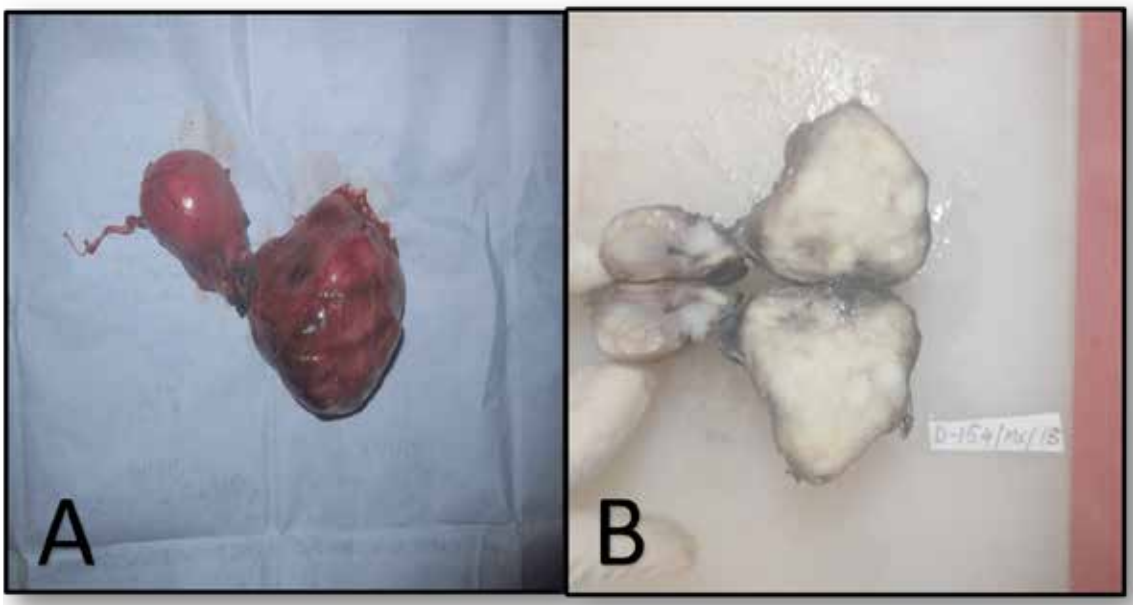

Figure 3: 3a): Surgically removed mass in toto; 3b): Formalin fixed gross tissue reveals the smaller buccal and larger parapharyngeal masses connected by the capsule.

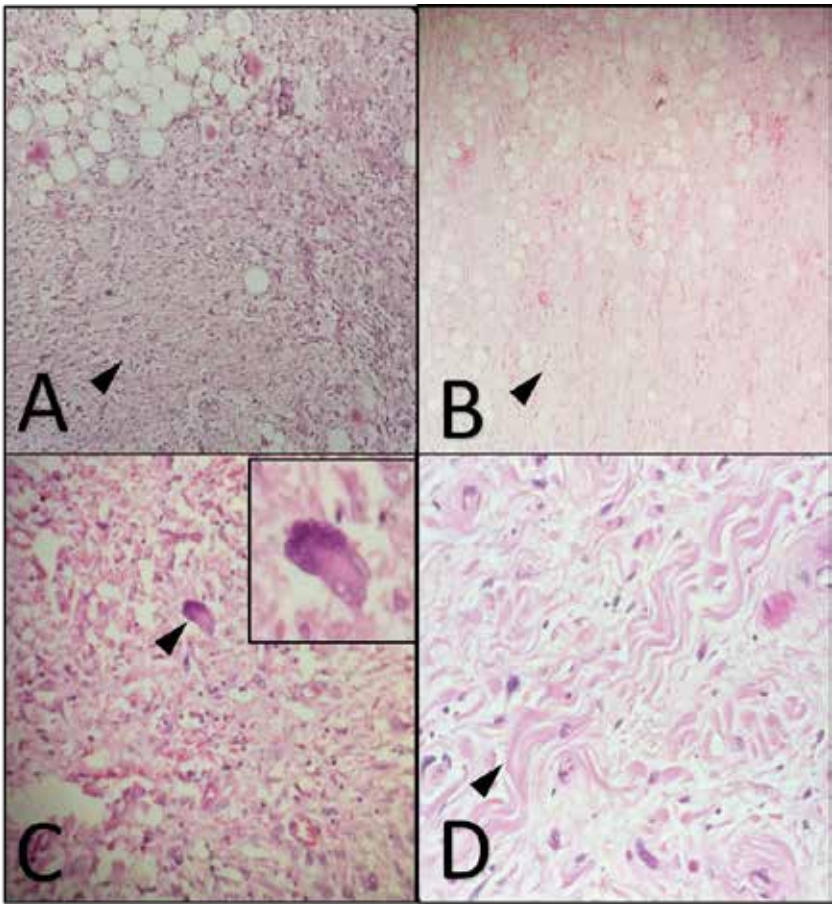

Figure 4: 4a): A photomicrograph reveals mature adipose tissue divided into lobules with areas of spindle cells (10X, H/E, arrowhead); 3b): interspersed myxoid changes in the stroma (10X, H/E, arrowhead); 3c): floret giant cells in the stroma (10X, H/E; arrowhead; inset: 40X, H/E); 3d): glassy, homogenized ropy collagen fibers (40X, H/E, arrowhead).

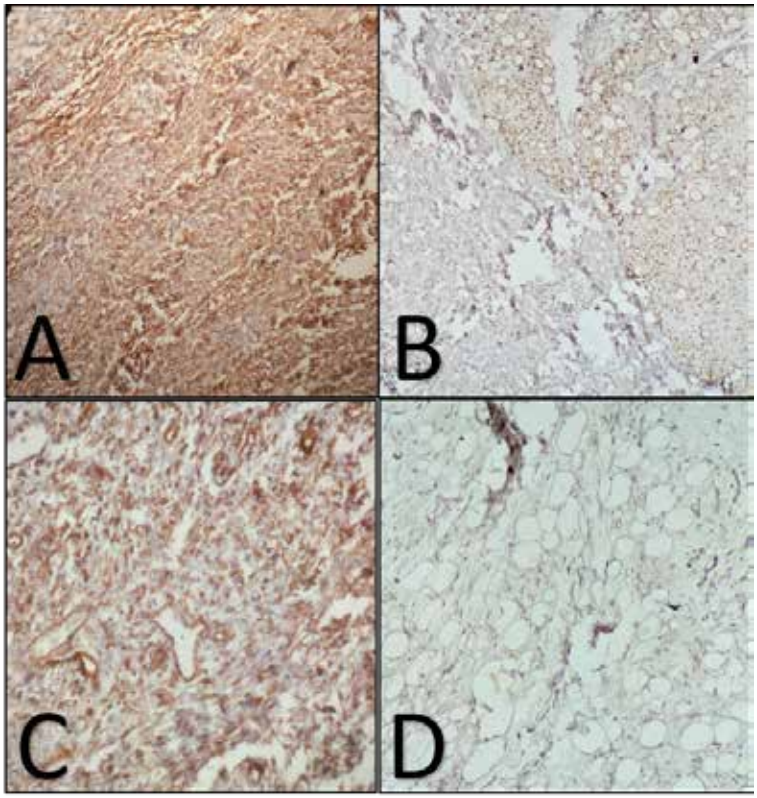

Figure 5: 5a): A photomicrograph reveals on IHC analysis spindle cells were strongly immunoreactive for CD34(10X, CD34); 5b): Spindle cells were negative but adipocytes show positivity for S100 (10X, S100); 5c): Spindle cells were strongly immunoreactive for vimentin (10X, Vimentin); 5d): Spindle cells and adipocytes were negative for desmin staining (10X, Desmin).

Table 1: Result of immunohistochemical analysis

IMMUNOHISTOCHEMICAL ANALYSIS		
MARKER USED	**POSITIVE CELLS**	**NEGATIVE CELLS**
S100	Adipocytes	Spindle cells
CD34	Spindle cells	Adipocytes
Vimentin	Spindle cells	Adipocytes
Desmin		Spindle cells, adipocytes

DISCUSSION

Spindle cell lipomas are infrequently seen in the oral cavity. The tongue is the most commonly affected oral site, followed by the floor of the mouth and buccal mucosa.[1] After our extensive literature search in indexed journals for this pathology since 1984 till date revealed only 45 cases with a mean age distribution of 55 years. SCL usually presents as solitary swelling besides only two cases reported with multiple presentation of SCL intraorally.[6-8] Cases with oral SCL were typically older adults with a male predilection whereas our case reports in an adolescent.[4] Clinically SCL is presented as a painless mass or swelling, often with a history of slow growth over an extended time period. They are most often solitary, though bilateral SCLs of oral cavity are also reported.[8] On gross examination, they are well circumscribed neoplasms with a firmer consistency than ordinary lipomas and cut surface may vary from yellow to grey-white. Histologically SCL consists of a mixture of mature adipocytes and bland spindle cells. The two components may be present in variable proportions with either one predominating.[1-5] These spindle cells were found to be associated with bundles of collagen having a wirey or ropy appearance. A myxoid stromal matrix is also a prominent feature in the histology of this tumor.[4]

Spindle cell lipoma and pleomorphic lipoma, share the same clinicopathologic and genetic features, except for the presence of enlarged multinucleated stromal giant cells. IHC study using S100, CD34, vimentin and desmin gives further more diagnostic advantage in confirming SCL. The spindle cells are found consistently positive for CD34.[1] Though this marker has limited role in confirming the diagnosis of spindle cell lipoma as CD34 expression is found positive in various other lipomatous and nonlipomatous soft tissue tumors.[1] Consequently immunoreactivIty for S100 protein is negative for spindle cells whereas positive for adipocytes, similar with the result in our case.

Similar to SCLs, well differentiated liposarcomas (WDL) are also present clinically as painless, slow growing masses.[3] Histologically, WDL can exhibit a spindle cell component that can lead to a histological similarity with SCL.[2,3] Though the histological features like variation in adipocyte size and shape, fibrous septa with atypical stromal cells exhibiting nuclear hyperchromasia, and the presence of more than an occasional lipoblast distinguish WDL from SCL. Lipoblasts, represented by multivacuolated and pleomorphic adipocytes, are absent in SCL, but are a component of WDL.[2,3] SCL with a predominant myxoid stroma, can mimic

myxoid liposarcoma. Myxoid liposarcoma histologically presents with rich capillary network and small signet ring lipoblasts which is not seen in SCL.[2] If the myxoid background is prominent, a pleomorphic adenoma with lipomatous change or metaplasia can be suspected but the lack of a plasmacytoid appearance, epithelial and myoepithelial elements, along with immunoreactivty with S-100 protein can help in ruling it out.[2]

Other pathological entities in the differential diagnosis of SCL are non-lipomatous neoplasms with the presence of spindle cells and adipose tissue, include fat-forming solitary fibrous tumor (lipomatous hemangiopericytoma), mammary-type myofibroblastoma, and cellular angiofibroma.[2,3] The fat-forming solitary fibrous tumor is composed of both adipocytes and CD34 positive spindle cell elements but the presence of a prominent branching type vasculature is a characteristic feature of fat-forming solitary fibrous tumor which is not prominent in SCL. In case of mammary-type myofibroblastoma, a more prominent intersecting fascicles of spindle cells and thickened, hyalinzsed bands of collagen are observed as compared to SCL.[2,10] Both tumors express CD34 but desmin positivity is more in mammary-type myofibroblastoma.[10] Histologically angiofibroma consisting of adipocytes along with the spindle cell component has a similarity with SCL. However, it lacks the conspicuous, thick walled, hyalinized vessels characteristic of angiofibroma. The collagen present in SCL is more eosinophilic and ropy than angiofibroma, which is more delicate and wispy in appearance. One rare differential diagnoses to be considered is the neurofibroma which in oral cavity is mostly seen on palate. The fusiform component in neurofibroma which is CD34 negative exhibits a wavy nuclei which is not seen in spindle cells of SCL.[2]

The exact origin of the fusiform component in SCL is uncertain, however, ultrastructural studies suggest that these spindle cells are modified fibroblasts originated from mesenchymal cells, which have lost their ability to differentiate into lipocytes but are capable of collagen synthesis. Hence these spindle cells show immunoreactivty to CD34, and are negative to alpha-smooth muscle actin, Factor VIII, cytokeratin, and S-100.[3]

The treatment of choice for spindle cell/pleomorphic lipoma is a complete local excision and its recurrence is extremely rare.[5] The present case has been under regular follow up for 14 months and has shown no signs of recurrence.

In English language literature till 2018, only 13 cases reported on primary parapharyngeal space lipomas in adults and so far only one case of pleomorphic variant of SCL in PPS has been reported (2001).[9] The multiple presentation or intercommunication of parapharyngeal lipomas are seen in many cases in which they may extend to base of skull, vocal folds, parotid gland, foramen transversarium encasing the vertebral artery, thyroid lobe thyroid cartilage or even to the tip of epiglottis and extending into the carotid space. But to our knowledge no case has reported an interspace communication of a parapharyngeal lipoma into buccal space.

CONCLUSION

We present a rare case of pleomorphic variant of SCL at multiple locations (parapharyngeal and buccal spaces) with an intercommunication by its capsular material. The present case, to our knowledge is unique by the youngest age of occurrence of SCL reported so far in the literature and the second case of pleomorphic SCL in parapharyngeal space. The histologic picture shows a range of variations and the observation of morphological features is important in distinguishing SCL from other fusiform tumors.

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