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Case Report

Buccal space malignant solitary fibrous tumour: a case report

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ABSTRACT

Solitary fibrous tumour (SFT) occurs rarely in extrapleural sites such as head and neck region. Head and neck region is affected in 16% cases occurring usually in orbit and paranasal region and are benign variants. Though usually benign, Malignant SFT in buccal space is extremely rare and comes with challenges in diagnosis and management. We report a rare case of Malignant SFT in the buccal space in a 70-year-old- gentleman who presented with slow growing mass in left cheek. Patient was treated with surgical excision and adjuvant radiotherapy and is on regular follow up. Malignant SFTs are difficult to diagnose due to lack of specific histological pattern but certain IHC and genomic markers can help. Buccal space malignant SFT can be managed by wide local excision and adjuvant radiotherapy in cases of large tumour size and margin positive resection.

Keywords: Solitary fibrous tumour, Malignant SFT, Buccal space lesion, Hemangiopericytoma

INTRODUCTION

Solitary fibrous tumour (SFT) was first described by Klemperer and Rabin as a rare spindle shaped tumour originating from the pleura in 1931.[1] Various terms were used to describe pleural SFT like fibrous mesothelioma, benign mesothelioma, localized mesothelioma, subpleural fibroma, and localized fibrous tumor of the pleura.[2]

SFTs constitute 5% of all sarcomas and up to 16% of them are found in head and neck region. Most of them are found within the orbit, and the most common variant is benign.[2] For many years, hemangiopericytomas (HPC) and SFT were considered as different entities. However, due to clinical, histological and immunohistochemical resemblance, most of the lesions previously diagnosed as HPC are now better defined as SFT.[5]

SFTs in buccal space present as mass lesion with its diagnosis on clinical grounds and needs a biopsy with lesion's microscopic appearance and characteristic immunohistochemical staining for CD34 determining its diagnosis. SFTs are now known to be associated with the recurrent gene fusion NAB2-STAT6 on chromosome 12q13 although no clear association between the prognosis and the genetic aberration have been identified.[9,10]

Newer modalities for diagnosis either by IHC stain for STAT6 or an RTPCR for NAB2-STAT6 fusion gene have been identified.[8,10]

Although 10-15% cases have been described to be malignant, such an occurrence of a malignant SFT in the buccal space is extremely rare in literature.

Surgery is main modality of treatment in head and neck SFTs but due to proximity of vital structures and the resultant morbidity, wide excision is possible in selected localised cases. Radiotherapy has been used as an adjuvant modality of treatment which can be used in high-risk cases.

Here we describe a case of Malignant SFT (MSFT) arising from left buccal space and its management with

surgical excision and adjuvant radiotherapy and with patient now being on regular follow up for 6 months.

CASE REPORT

A 70-year-old gentleman with no co-morbidities presented with slow growing swelling over left cheek since 4 years (Figure 1). On examination with bidigital palpation a 4x4cm well defined, lobulated, firm, non-tender, mobile swelling over left buccal space with overlying skin and underlying buccal mucosa intact and free (Figure 2). CECT revealed an oval shaped heterogeneously enhancing predominantly solid lesion in left buccal space of 4.2×3.4×3.9 cm with preserved fat planes with surrounding structures.

Figure 1: Clinical picture of left cheek swelling.

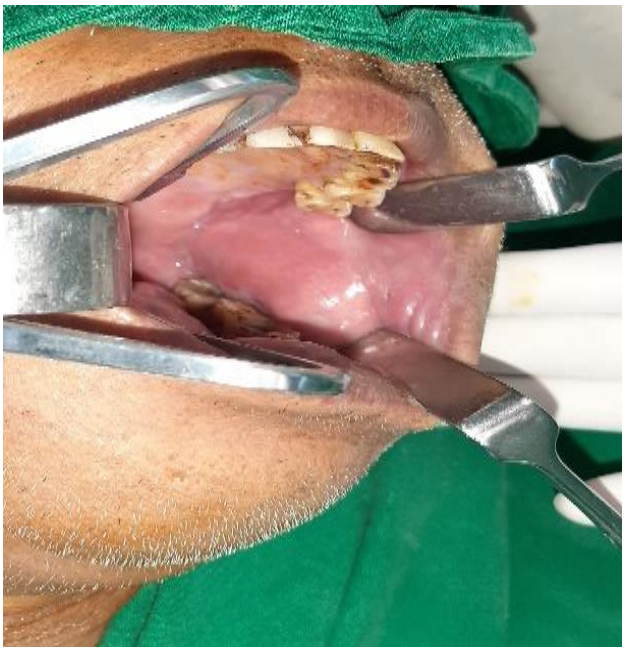

Figure 2: Clinical picture of intraoral aspect of left buccal space swelling.

USG guided FNAC from the tumour reported as monomorphic adenoma of salivary gland or dermal analogue of basal cell adenoma

He underwent enucleation of the lesion through intraoral route; intraoperatively two well defined lobulated lesions in left buccal submucosal plane were noted (Figure 3 and 4).

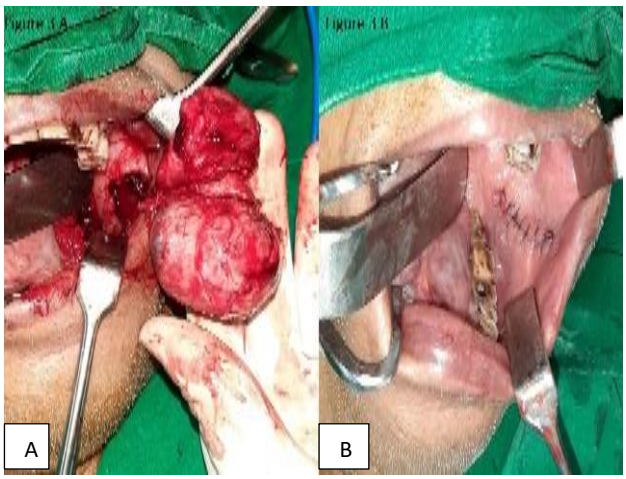

Figure 3 (A and B): Intraoperative picture of intraoral excision of bilobed left buccal space lesion and primary closure of surgical incision.

Figure 4: Excised surgical specimen.

Histopathology revealed-malignant SFT, encapsulated, comprising spindle shaped cells with intervening collagen in palisade appearance (Figure 4). Cells exhibited mild to moderate pleomorphism, increased mitotic figure>4/10 hpf. Tumour focally infiltrated surrounding capsule. Immunohistochemistry-CD-34 positive (Figure 6A), vimentin-strong and diffuse positive, STAT-6 nuclear positive (Figure 6 B, 6 C), negative for S100, p63, SMA and Ki-67-35% (Figure 6 D).

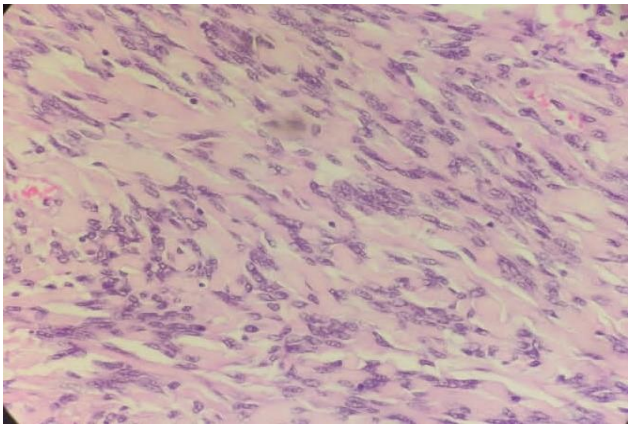

Figure 5: Histopathological examination slide (40X).

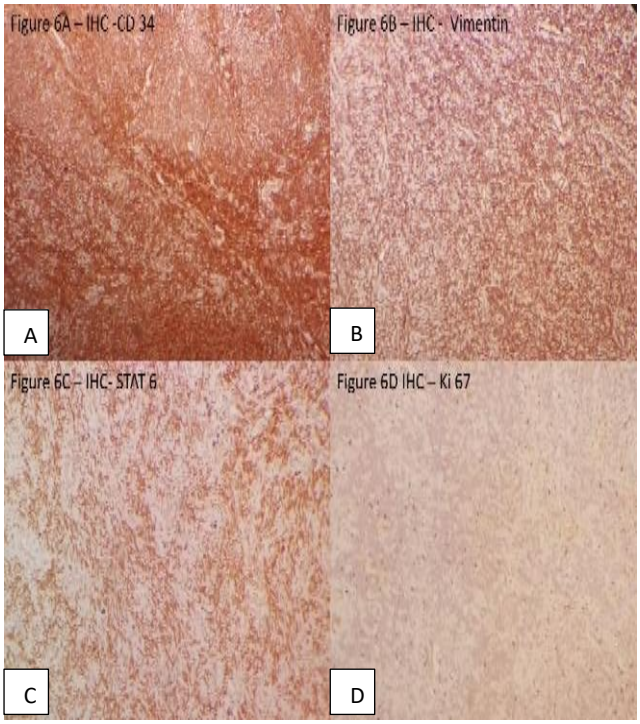

Figure 6 (A-D): Immunohistochemistry, IHC-CD 34, IHC-vimentin, STAT 6 and IHC-Ki67.

A multidisciplinary tumour board decision was made to subject the patient to adjuvant radiotherapy. Patient received External beam. Patient is now on regular follow up for 6 months with no signs of recurrence.

DISCUSSION

SFT were first described in 1931 as to be originating from pleura and they roughly account for 5% of all sarcomas.[1,2]

It is uncommon in head and neck accounting for 16% of all cases. Found within the orbit and paranasal sinus and mainly are benign.[6] First head and neck region SFTs was described by Witkin et al in 1991 where 6 cases occurring in nasal cavity in adult patients were reported. None of them were described to be malignant.

Histologically, SFT has been described as a spindle cell tumor with a patternless architecture, hemangiopericytoma-like branching vascular pattern and strong CD34 immunoreactivity.[4] SFTs of extra-pleural origin have been diagnosed with increasing frequency in recent years as the result of improved methods of pathologic examination. Under the world health organization (WHO) classification of tumors of soft tissue and bone in 2013, hemangiopericytomas (HPC) and SFTs of the soft tissues are regarded as features of the same entity in the soft tissue fascicle.[5]

Diagnostic criteria for SFT included a cytologically bland spindle cell lesion with variable cellularity and focal dense collagenization with diffuse, strong CD34 and CD99 reactivity, while the diagnosis of Malignant SFT (MSFT) is dependent on combining factors such as mitotic activity, cellularity, presence of hemorrhage and necrosis.

Microscopically, MSFTs are usually hypercellular lesions, showing at least focally moderate to marked cytological atypia, tumor necrosis, numerous mitoses (>4 mitoses per 10 HPF) and/or infiltrative margins.[7]

Our case had mitoses of >4/10 hpf and also focally infiltrating the surrounding capsule.

IHCs commonly used in SFT include CD34, Bcl-2 and CD99 with specificity of 97.2%, 91% and 72% respectively. Moreover, SFT is notably negative for other well-known markers that may also contribute to the correct diagnosis, like cytokeratin, desmin, epithelial membrane antigen (EMA), α-SMA and S-100 protein, and these markers are frequently used to endorse the diagnosis of SFT.[8,9]

Genomic inversion found to occur in SFTs which causes NAB2-STAT6 fusion into a common direction of transcription. Consequently, STAT6 converts NAB2 into a transcriptional activator of EGR-1 (early growth response protein-1), a mitogenic pathway inducing neoplastic progression.

IHC for STAT6 has specificity of up to 90%-99% and RTPCR for NAB2-STAT6 fusion up to 90% in SFTs.[8,10]

Malignant SFT of oral cavity have an overall favourable prognosis with 14 cases described in literature, one patient had metastases and one had local recurrence.[8] Till date only 3 cases of malignant SFT of buccal space have been described, with ours being the 4th case (Table 1).

Table 1: Clinicopathological characteristics of malignant SFT of buccal mucosa.

Cases	Age (Years)	Sex	Size (cm)	Mitosis (Hpf)	Necrosis	Treatment	Recurrence	Follow up (Months)
1	62	Male	4.1	NA	NA	Surgery	No	41
2	27	Male	2	6/10	No	Surgery	Yes	39
3	65	Female	NA	>4/10	NA	Surgery	No	18
4 (Our case)	70	Male	4.2	>4/10	No	Surgery + RT	No	6

NA-Not available.

Surgery with wide healthy margins is recognized as the gold standard, but in areas such as orbit, nasal cavity a wide margin is difficult to attain. Factors that predispose to local recurrence in non-head and neck SFTs are a tumor diameter larger than 10 cm, the presence of a malignant component and positive surgical margins.[11]

Adjuvant radiotherapy has been shown to improve local control with 5-and 10-years control rates of 100% when used in malignant SFTs >5 cm in size or when surgical margins are inadequate.[12] Our patient underwent adjuvant radiotherapy as preoperatively he was diagnosed to have a benign lesion and underwent enucleation with a combined tumour size was 7.5×4×3 cm.

Recurrence rate for MSFT of head and neck has been described in case series by Yang et al were three out of 9 patients developed locoregional and distant metastases ranging from 4 months post-surgery to 7 years, SFT at other sites have been described to recur longer than 5 years duration hence a regular follow up needed to detect such recurrence.[7]

CONCLUSION

It is difficult to diagnose malignant SFT in buccal space preoperatively as it lacks specific histological characteristics in biopsy. Thorough evaluation is needed to identify malignant nature to optimise the treatment thus avoiding recurrence. A long term follow up is required as late recurrence is observed in some cases.

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