



Synovial Sarcoma of Buccal Mucosa: A Case Report

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Abstract

Synovial Sarcomas are distinct malignant soft tissue tumors of pluripotent mesenchymal cells. In head and neck SS occurs rarely comprising only 2.5% to 3.5%. SS is rare in oral cavity. We report a case of synovial sarcoma of the buccal mucosa in a 23-year-old female patient treated by right buccal mucosa wide excision with supraomohyoid neck dissection and reconstruction with Anterolateral thigh flap followed by 3D conformal external beam radiotherapy. The prognosis of synovial sarcoma is difficult to formulate. Complete excision of the tumor with wide surgical margins is the mainstay in the treatment of SS. A multimodal approach consisting of wide local excision of tumor followed by postoperative radiation and chemotherapy is often recommended as complete excision of intraoral tumors is not always possible. Incomplete surgical resection & local recurrence are associated with increased morbidity and mortality.

Keywords: Head and Neck Synovial Sarcoma; Oral cavity; Buccal mucosa; Multimodality treatment

Introduction

Synovial sarcomas are distinct malignant soft tissue tumors of pluripotent mesenchymal cells. SS primarily occurs in the paraarticular regions of the extremities, in close association with the bursa, tendon sheaths and joint capsules. Synovial sarcoma accounts for 8% to 10% of all sarcomas [1]. Approximately 90% of SS have the chromosomal translocation (X; 18), which results in the formation of a fusion product between the synaptotagmin1 gene, SYT1, on chromosome 18 and the SSX family member 1 gene, SSX1, or SSX family member 2 gene, SSX2, on chromosome [2].

In head and neck SS occurs rarely comprising only 2.5% to 3.5%. It was first described in the head and neck in 1954, when Jernstrom reported a case of synovial sarcoma of the pharynx. Head and neck SS have a hematogenous spread hence greater potential for both distant and regional metastasis, as compared to other SS. The most common site for HNSS has been reported in soft tissues of the neck and upper aerodigestive tract. HNSS is more common in middle aged males [3].

SS is rare in oral cavity. HNSS most commonly presents as a painless mass & may be associated with bleeding, dysphagia, hoarseness and odynophagia. The treatment of HNSS comprises of wide local excision plus adjuvant radiation and/or chemotherapy. We report a case of synovial sarcoma of the buccal mucosa in a 23-year-old female patient.

Case Presentation

A 23-year-old female reported to the Department of Head and Neck Oncology with the chief complains of swelling on right side of face since 2 months. Initially swelling was smaller in size which gradually increased to the present size; there was no associated pain or decrease in mouth opening. Patient had family history of Neurofibromatosis. There was past history of monophasic synovial sarcoma of right buccal mucosa treated by wide excision of buccal mucosa in 2/6/2020 without any adjuvant treatment. IHC revealed neoplastic cells expressing vimentin, TLE-1, CD-99, CK, Desmin, Ki67 with no reactivity for S 100, SOX 10, GFAP, HMB 45, P63, CK5/6, CD34. On general physical examination, the patient was moderately built and well nourished. All the vital signs were within the normal limits.

Extra oral examination revealed facial asymmetry with a diffuse swelling on right side measuring 5 cm × 4 cm in dimension. Swelling was extending from 1.5 cm below the lower eyelid superiorly to the lower border of the mandible inferiorly. Anteriorly 1 cm from the lip commissure extending posteriorly till junction of angle and ramus of mandible. Tethering of overlying skin was noted.

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Figure 1: Right buccal mucosa synovial sarcoma.



Figure 2: Endophytic lesion on right buccal mucosa.

Swelling was non tender on palpation and firm in consistency.

Intraoral examination revealed an endophytic growth measuring approximately 3 cm × 4 cm on the right buccal mucosa involving upper and lower gingivobuccal sulcus, going into the RMT. The lesion extended anterior posteriorly from 2nd premolar tooth to RMT region. The growth was tender on palpation and firm in consistency.

Investigations included complete hemogram, PET CT Scan, and biopsy. PET CT revealed an intensely FDG avid enhancing globular lesion arising from masseter muscle. (SUVmax 49.6 g/ml; 4.6 cm × 2.9 cm × 3.7 cm). It abuts the periosteum of mandibular ramus with no obvious erosion. FDG avid lytic lesion is seen in anterior part of right mandibular ramus (SUVmax 6.2 g/ml). FDG avid enhancing nodule is seen in the left erector spinae muscle at the level of T9 vertebra (SUVmax 3.5g/ml; 1.6 cm × 1.4 cm × 2.4 cm). Low grade FDG uptake seen in the pleural based nodule in the left lower lobe along the costal pleura (SUVmax 4.1g/ml; 3.9 cm × 2.3 cm × 3.5 cm). Biopsy and

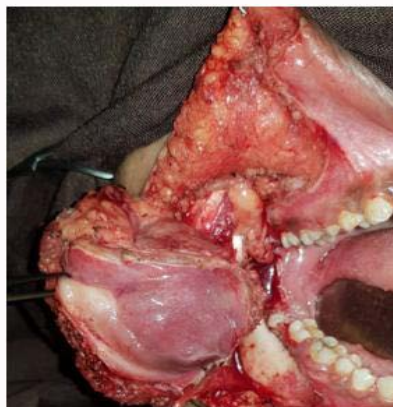


Figure 4: Full thickness wide local excision of right buccal mucosa.

IHC from right buccal mucosa was suggestive of recurrent biphasic Synovial Sarcoma. USG guided biopsy from left erector spinae muscle revealed spindle cell carcinoma which on IHC examination was suggestive of neurofibroma; spindle cells were immunopositive for S-100 and SOX 10 and negative for cytokeratin, EMA, SMA, Desmin and CD34. CT guided biopsy from pleura showed no specific lesion so a provisional diagnosis of neurofibromatosis I with recurrent SS was made.

Treatment consisted of right buccal mucosa wide excision with supraomohyoid neck dissection and reconstruction with anterolateral thigh flap followed by 3D conformal external beam radiotherapy.

Discussion

Synovial sarcoma is a malignant soft tissue tumor arising from a pluripotent mesenchymal stem cell capable of differentiating into cells with epithelial and fibroblast-like features [4], which usually occurs at the extremities of young adults. It is rare in the head and neck region. The pharyngeal and the hypopharyngeal areas are most commonly affected [4] in the head and neck region [5]. Although synovial sarcoma may occur at any age, it is most commonly found in adolescent and young adults with a sex ratio of about 1.2:1 M:F. At diagnosis, patients have a mean age of 31 to 36 years with 60% to 70% of patients under 40. It's a slow growing tumor which expands locally and forms a pseudo-capsule thus compressing the normal surrounding tissue. The most typical presentation of synovial sarcoma is of a painless mass but some authors report that up to 40% are painful.

Differential diagnosis must be made from fibrosarcoma, leiomyosarcoma, spindle cell sarcoma, malignant schwannoma and adenocarcinoma. SS is classified as monophasic or biphasic depending

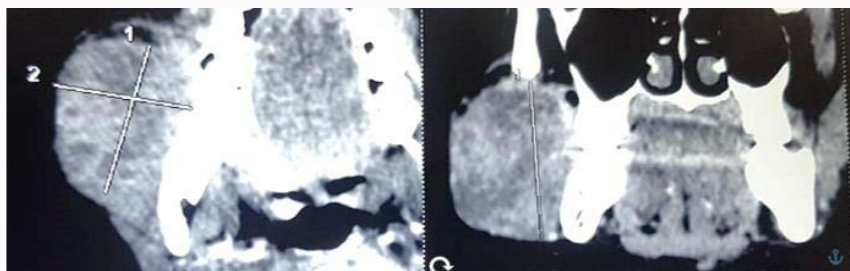


Figure 3: Axial and Coronal section showing an intensely enhancing 47.9mm × 52.4mm × 39.6mm soft tissue lesion is seen arising from right masseter muscle involving the superior and inferior Gingivo buccal sulcus. It abuts the periosteum of mandibular ramus, but with no obvious erosion. Paramandibular soft tissue covering entire height of adjacent body of mandible.



Figure 5: Excised specimen.



Figure 6: 2 Months follow up.

| IMMUNOHISTOCHEMISTRY REPORT | |
|-----------------------------|--|
| IHC MARKER(S) | RESULT |
| Vimentin | Immunoreactive in neoplastic cells (score 4+) ✓ |
| TLE-1 | Immunoreactive in neoplastic cells (score 4+) ✓ |
| CD99 | Immunoreactive in neoplastic cells (score 4+) ✓ |
| CK | Immunoreactive in neoplastic cells (Focal, score 1+) ✓ |
| Desmin | Immunoreactive in neoplastic cells (Focal, score 1+) ✓ |
| S-100 | Non-immunoreactive in neoplastic cells |
| SOX-10 | Non-immunoreactive in neoplastic cells |
| GFAP | Non-immunoreactive in neoplastic cells |
| HMB-45 | Non-immunoreactive in neoplastic cells |
| p63 | Non-immunoreactive in neoplastic cells |
| CK5/6 | Non-immunoreactive in neoplastic cells |
| CD34 | Non-immunoreactive in neoplastic cells |
| KI-67 | Immunoreactive in 60-65% neoplastic cells (score 4+) ✓ |

Clinical Information: 4/10 right cheek swelling. 4/10 pre-meal excision on 24.2020. HP report (MPSGAC) 11.6.2020 - 8th from right side cheek swelling. soft tissue sarcoma. IHC report (MPSGAC) 10.6.2020 - soft tissue sarcoma ? myxoid fibrosarcoma liposarcoma ? MPNST. CT scan of neck & face 29.5.2020 - Enhancing soft tissue lesion 6.8 x 3.8 x 4 cm involving right che subcutaneous plane with internal non-enhancing necrotic area. Skin & mandible free. Posteriorly causes scalloping of masseter in possible loss of fat plane. Cytology (MPSGAC) 27.5.2020 - FNAC from right sided cheek swelling. 5/10 borderline neoplasm (liposarcoma) ? soft tissue tumor ? epithelial tumor.

Reference HP No.: Outside H/1212/20

Testing Performed on Block Number(s): 4051/20.21

Specimen: Right side cheek swelling.

HP Diagnosis: Soft tissue sarcoma ? Angiosarcoma ? myxoid fibrosarcoma ??? myxoid liposarcoma

Impression: Monophasic Synovial Sarcoma

Figure 7: Histopathology and IHC Report of first surgery.

upon the presence of spindle cell component or an epithelial and spindle cell component both, respectively. Additionally a “poorly differentiated” type of SS was described by Enzinger and Weiss which shows more aggressive behavior [6]. Dystrophic calcification and calcospherites are characteristic findings in synovial sarcoma.

Expression of Cytokeratin (97%) and EMA (69%) is seen in all SS, although to a variable extent. Synovial sarcomas usually express CK7 and CK19/32; however, this immunoreexpression may be only focal in the monophasic and poorly differentiated synovial sarcomas. S-100 protein is expressed in 30% of tumors while CD99 is seen in 67% of all SS. Expression of bcl-2 protein is common in SS. Epithelial and spindle cells of biphasic, monophasic, and poorly differentiated tumors express Calretinin and is of diagnostic significance in SS. Spindle cell components have been described as being more often positive than the epithelial cells [7,8]. According to some studies, TLE-1 expression, is a sensitive and specific marker for SS, and could be as high as 90% [9,10].

The prognosis of synovial sarcoma is difficult to formulate. According to the literature HNSS have a relatively good prognosis compared with other SS. The 5 year OS rates has been reported to be between 47% to 82% [3,11]. OS and PFS is poorer in larger tumors. Although most studies have utilized the 5 cm cutoff, Enzinger and Weiss believed that size influenced prognosis as patients whose tumors were less than 4 cm in diameter seemed to do better than those with larger tumors. The influence of histological differentiation on the prognosis is unclear. Schmookler et al. [13] found no association between the outcome and pathological features of the synovial

sarcoma on head and neck. However, Enzinger and Weiss have correlated improved prognosis with degree of differentiation of the tumors and calcification within the tumor. The prognosis of synovial sarcoma is adversely affected by tumor location, size, patient’s age, surgical procedure, degree of differentiation [12-14]. It is believed that: 1) age <60 years, 2) appropriate surgical resection, 3) total tumor dimension <5 cm, 4) extensive calcification, 5) a high degree of tumor differentiation, extensive hemorrhagic necrosis and high mitosis index, 6) tumor without distant metastasis, will lead to good prognosis [15-17]. The presenting case is biphasic with epithelioid and spindle cell morphology. Maximum tumor dimension is about 5 cm, Necrosis (approximately 5%) present. Mitotic activity is >40/10 hpf.

Complete excision of the tumor with wide surgical margins is the mainstay in the treatment of SS. Achievement of negative margins is important in avoiding local recurrence [18]. A multimodal approach consisting of wide local excision of tumor followed by postoperative radiation and chemotherapy is often recommended as complete excision of intraoral tumors is not always possible. As compared to SS originating at other sites, the metastatic potential (29.2%) and recurrence rate (20.8%) of the oral tumors is low. Multiple recurrences were not rare. Most commonly, SS metastases to the lung, followed by the lymph nodes and bone marrow [19]. Tumor recurrence is typically manifested in the first 2 years after initial therapy.

According to the Yang’s report, after observation of 21 patients with synovial sarcoma of the head and neck, it was found that postoperative chemotherapy slightly prolonged the time for the occurrence of distant metastasis but showed no significant difference for the overall survival rate or local recurrence [20]. In general, chemotherapy in HNSS should be given only in presence of poor prognostic factors or an unfavorable site of presentation.

Conclusion

Head and neck sarcomas are rare. Incomplete surgical resection & local recurrence are associated with increased morbidity and mortality. Wide excision with post-operative adjuvant therapy

remains the mainstay of treatment of SS. The prognosis and survival can be improved with appropriate early diagnosis and treatment of patients.

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