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ORIGINAL RESEARCH

Synovial Sarcoma of the Floor of the Mouth: A Rare Clinical Entity with Review of Literature

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ABSTRACT

Background: Head and neck synovial sarcoma (SS) amounts to 6.8% of all total body SS, with high incidences in the hypopharynx, postpharyngeal region and the parapharyngeal space. Also, incidences of SS in the tongue, soft palate, mandible, buccal mucosa, floor of the mouth, lungs, thymus, trunk and posterior peritoneum are reported.

Case report: We report a case of 12-year-old girl with swelling in the floor of the mouth for 2 months. The patient had difficulty in speech, tongue movements, poor oral hygiene due to food residue collecting around the tumor in the floor of the mouth. A smooth 4 × 4 cm globular mass arising from the floor of the mouth with the fine needle aspiration cytology (FNAC) as spindle cell neoplasm. Wide excision and immunohistology confirmed it as calcified synovial sarcoma, hence postoperative chemotherapy was given. There is no recurrence for the past 3 years.

Conclusion: Synovial sarcoma is a rare tumor in the oral cavity, which is slow growing and of variable aggressiveness. Immunohistological analysis confirming the diagnosis and wide surgical excision with adjuvant chemotherapy is the modality preferred in these cases. Recurrence is common in the first 2 years of therapy but can occur later also.

Keywords: Floor of the mouth, Metastasis, Recurrence, Synovial sarcoma.

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INTRODUCTION

Synovial sarcoma (SS) is a rare malignant neoplasm seen in the capsules of joints, articular tendons and one of the most common malignant soft-tissue sarcomas in children and adolescents.[1,2] They form 5.6 to 10% of all soft-tissue sarcomas and the fourth most common soft-tissue malignancy after malignant fibrous histiocytoma, liposarcoma and rhabdomyosarcoma.[3,4]

Controversy still exists at its synovial tissue of origin, as its now universally accepted they originate from undifferentiated or pluripotent mesenchymal cells both epithelial and mesenchymal differentiation capacity.[5]

Head and neck SS amounts to 6.8% of all total body SS, with high incidences in the hypopharynx, postpharyngeal region and the parapharyngeal space.[2,6] Also, incidences of SS in the tongue, soft palate, mandible, buccal mucosa, floor of the mouth, lungs, thymus, trunk and posterior peritoneum are reported.[3] Synovial sarcoma appears in the third decade in young adults with incidences twice commoner in males than females.[3]

CASE REPORT

We report a case of 12-year-old presented with swelling in the floor of the mouth for 2 months. The patient had complaints of difficulty in speech, tongue movements, poor oral hygiene due to food residue collecting around the tumor in the floor of the mouth. A smooth 4 × 4 cm globular mass arising from the floor of the mouth mobile not fixed to the mandible was seen with firm hard induration surrounding the mass lifting the tongue dorsally. Right-sided level 2 lymph node was hard, mobile and nontender. A provisional diagnosis of spindle cell neoplasm on fine needle aspiration cytology (FNAC) with the node being negative of stage $T_2N_0M_0$ was made. Contrast-enhanced computed tomography (CT) images showed calcified lesions in the floor of the mouth with no infiltration into the mandible (Fig. 1).

Wide excision transorally was done and the recovery was uneventful. The histopathological examination revealed calcifying synovial sarcoma with marker studies showing positive for epithelial membrane antigen (EMA), BCL-2, negative for SMA, desmin and CD34. The patient was given two cycles of adjuvant chemotherapy. No recurrence was seen in the past 3 years (Fig. 2).

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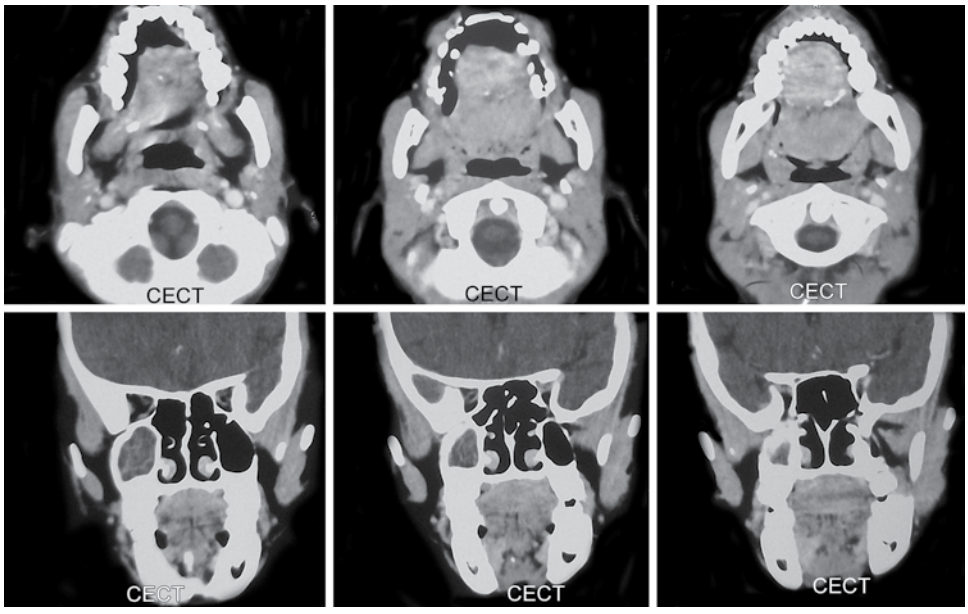

Fig. 1: Axial and coronal cuts showing calcified mass in the floor of the mouth

DISCUSSION

Synovial sarcoma (SS) is very rare malignant soft-tissue tumor, occurring in the head and neck region with aggressive course and metastatic potential.[1] As the incidence is lesser reported in the floor of the mouth, ambiguity about it being malignant, primary and metastatic spindle cell tumors of the oral cavity still exist.[7] In the head and neck, SS present as dysphagia, hoarseness or headache depending on the origin and expansion of the planes of the tumor.[8] In other parts of the body, they present as slow-growing palpable mass mimicking a benign course.[9] They range from smaller to a 15 cm lesion, the superficial detected earlier while the deeper goes undiagnosed for longer time.[10]

According to the RMH classification, all the tumors of low grade below 15 cm and high grade below 10 cm were staged below III.[1] All SS with nodal metastasis as stage IVA and with distant metastasis as stage IVB.[1] Biphasic tumors, monophasic tumors, monophasic epithelial tumors and poorly differentiated (round cell) tumors are four subtypes described while monophasic epithelial type, monophasic spindle cell type and biphasic type with distinct epithelial and spindle cell components are three variants noted in literature.[11] All these tumors are regarded as carcinosarcoma of soft-tissue origin regardless of subtypes.[12]

Fig. 2: Good control 2 years after surgery and chemotherapy

On histopathological examination, SS shows a characteristic biphasic pattern with epithelial round cell component resembling carcinoma and a fibrosarcoma-like spindle cell component.[13] Generic transitions are seen between the epithelial and spindle cell lines.[3] Both epithelial and spindle cells are reactive with cytokeratin (CK) and epithelial membrane antigen (EMA), while only the spindle cells are positive for vimentin.[14] Vimentin, Bcl-2, CD99 and Mic-2 with enhanced cytoplasmic positivity suggests a poorly differentiated small cell variant synovial sarcoma.[4] Cytogenetic analysis shows a fusion gene (SYT-SSX), which is specific for SS.[14] A characteristic translocation t (X; 18) (p11.2;q11.2) is seen in SS in other parts of the body which is also seen in SS in the head and neck.[15]

Surgical excision with adequate negative margins is the optimal management in other parts of the body; while in head and neck, a combined therapeutic approach consisting of extensive radical local excision, postoperative radiation therapy and chemotherapy is often recommended.[1] Locoregional recurrence up to 60 to 90% in 2 years are reported without adequate surgical margins in SS all over the body.[16] No official data about the efficacy of radiotherapy and chemotherapy have been reported.[13] The tumors in the head and neck has a metastatic potential of 12.5 to 29.2% and recurrence rate of 20.8% which is lower compared to other sites.[6]

Metastasis is common to the lungs, lymph nodes and the bone marrow.[4] Recurrences are typically manifested in the first 2 years after initial therapy and also reported after 20 years.[1] As SS is an aggressive tumor, 36 to 80% 5-year survival rate and, 50% 10-year survival rate are reported.[17] Larger tumors more than 5 cm, tumor site, older patients, high-grade malignancies, high-mitotic activity, compromised margins of resection and metastasis makes the prognosis worse.[18] Contrast-enhanced CT images show enhanced solid masses, while MR images show a smooth margin well-defined mass with minimal infiltration.[19,20] Lesions appear homogeneous or heterogeneous masses depending on extent of hemorrhage or necrosis. Thirty percent of the tumors show calcifications which represent better prognosis.[19,20] On MR, they appear isointense to gray matter on T1-weighted images and is well defined yet heterogeneous with septations, hemorrhage, cysts, calcification or multilocularity but not specific for SS.[19,20]

Synovial sarcoma is often misdiagnosed as CECT, MR images show a benign smooth margin, cystic components and lack of aggressive infiltration.[20] So, we emphasize on an early diagnosis and aggressive management as recurrence and metastasis are common and variable.[20] Jernstrom et al first reported a case of head and neck SS in the pharynx.[21,22] Later, Hiroto et al reviewed 100 cases head and neck SS and reported the minimal incidence of cervical metastasis.[13] They reported a biphasic synovial sarcoma of the right tonsil with neck metastasis treated by wide excision and modified radical neck dissection.[13]

Park et al reported a case of SS located near the hyoid bone in the submental area which was predominantly cystic unlike the solid mass described in literature.[23,24] It was a variant carcinoma arising from thyroglossal duct remnant.[23,24] Wadhwan et al reported rarity in the orofacial region to the reason for delayed diagnosis.[3] They reported a rare biphasic synovial sarcoma in mandibular region occurring after hemimandibulectomy for ameloblastoma of the jaw.[6] Roth et al analysed 24 cases of SS of head and neck where he reported the mean age as 19 years, with slight male predominance with most of the tumors were round smooth lobulated swellings.[3,25]

CONCLUSION

Synovial sarcoma is a rare tumor in the oral cavity, which is slow growing and of variable aggressiveness. Immunohistological analysis confirming the diagnosis and wide surgical excision with adjuvant chemotherapy is the modality preferred in these cases. Recurrence is common in the first 2 years of therapy but can occur later also.

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