The "TROJAN HORSE" of a Dental Visit – Synovial Sarcoma

Abstract

The term "synovial sarcoma (SS)" is a histological error, a misnomer as it neither arises from nor differentiates toward synovium. Head and neck region is the most commonly affected region after extremities, representing 5% of all cases. This case report focuses to discuss a case of a SS that was diagnosed after an inadvertent root canal therapy. A 46-year-old male came to the outpatient department with a chief complaint of pain and swelling in his upper right back tooth region since 15 days. An ulceroproliferative mass of was observed protruding from the gingivobuccal sulcus from 11 to 15 tooth region obliterating the vestibule.

Keywords: *Paranasal sinus, rhabdoid, soft-tissue tumor*

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Introduction

The term Synovial Sarcoma is a histological error, a misnomer as it neither arises from nor differentiates towards synovium. Head and Neck region is the most commonly affected region after extremities, representing 5% of all cases. This case report focuses to discuss a case of a Synovial sarcoma that was diagnosed after an inadvertent root canal therapy.

Case Report

A 46-year-old male presented to the outpatient department with a complaint of pain and swelling in his upper right back tooth region since 15 days. The patient gave a history of pain in relation to 14 and 15 for which he underwent inadvertent root canal therapy, followed by apicectomy and extraction of 12, 3 weeks earlier at a local dental hospital. Posttreatment, the pain and swelling did not subside which lead the patient to revisit his local dental surgeon, where he was advised to undergo biopsy.

On extraoral examination, there was a significant facial asymmetry caused by an approximately 5 cm × 4 cm mass involving the right maxilla. No pulsations were detected, and overlying skin was normal in color. Intraorally, an ulceroproliferative mass of 4 cm × 5 cm in its greatest dimension was observed protruding from the gingivobuccal sulcus from 15 to 11 tooth region obliterating the vestibule. The lesional mass was reddish, firm in consistency, with ulcerated surface and tender on palpation. The lesional mass extended around the upper gingival area near right central incisor to right first premolar, completely masking the associated tooth surface and also obliterating the gingivo buccal sulcus. There was no evidence of tooth mobility and pus discharge [Figure 1]. The computed tomography scan of the paranasal sinus revealed an enhancing mass lesion in the right gingivobuccal sulcus and nasolabial fold extending into maxillary sinus, with destruction of anterior wall of the sinus, and erosion of the alveolar margin [Figure 2]. Right cervical lymphadenopathy was observed with 6–20 mm right Level IB and IIA. A provisional diagnosis of soft-tissue tumor was given. The differentials included nodular fasciitis, malignant peripheral nerve sheath tumor (MPNST), fibrosarcoma, and rhabdomyosarcoma.

An incisional biopsy was subsequently performed and subjected to histopathological examination. On microscopic examination, the hematoxylin and eosin-stained sections exhibited loose, hypercellular stroma with a mixture of cell types: predominantly proliferating neoplastic ovoid cells with spindle cells, tadpole-shaped cells, and rhabdoid type cells. The cells were large to intermediate in size with vesicular eccentric nuclei and eosinophilic cytoplasm, high mitotic activity, and hyperchromatism. Considering the aggressive clinical behavior, radiographic and histopathologic features,

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a diagnosis of rhabdomyosarcoma was provided and referred to local cancer hospital for further investigations and treatment. The patient underwent hemimaxillectomy with levels I–III cervical lymph node dissection, followed by adjuvant chemotherapy at local cancer hospital, and an excisional biopsy was submitted for histopathological examination.

The microscopic examination revealed coexistence of epitheloid cells and fibroblast-like spindle cells. Epitheloid cells were round to oval polygonal cells arranged in solid cords, nests or glandular structures with large, round/oval, hyperchromatic, vesicular nuclei, pale cytoplasm, and distinct cell borders. The fibroblast-like spindle cells were uniformly appearing, well-oriented, plump, with small amount of cytoplasm, oval dark-staining nuclei, solid, compact sheets similar to fibrosarcoma. The stroma also presented rhabdoid cells/racquet cells, neoplastic bone formation, cleft-like spaces – hemangiopericytoma like hyperchromatic and vesicular nucleus, increased nuclear-cytoplasmic ratio, mitotic figures, and hemorrhage was evident in the cells [Figure 3]. Immunohistochemistry revealed that the tumor cells were positive for cytokeratin and Bcl2, and negative for myoglobin, smooth muscle antibody (SMA), S100, CD 31, CD 56, HMB45, vascular endothelial growth factor (VEGF), and P63 [Figure 4]. A final diagnosis of "Poorly differentiated synovial sarcoma (SS) of rhabdoid variety" was made accordingly. The patient was treated with neoadjuvant chemotherapy initially, followed by surgical resection that incorporated hemimaxillectomy and followed by Radiotherapy. The patient subsequently underwent a complete whole-body positron emission tomography after 3 months, which revealed metastatic deposits in the lung and kidneys.

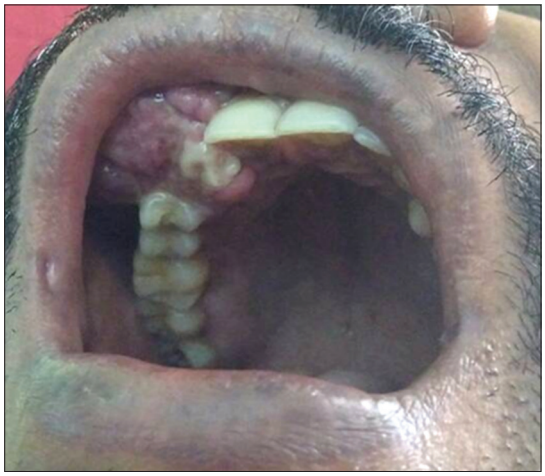

Figure 1: Intraoral picture of the lesion

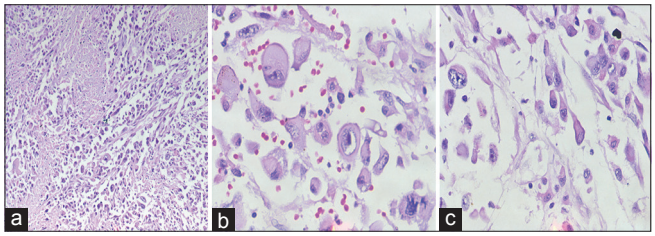

Figure 3: (a) ×10 view: Hypercellular areas presenting pleomorphic cells with hyperchromatic nuclei. (b and c) ×40 view: Large pleomorphic and tadpole shaped cells with ecentric, hyperchromatic vesicular nuclei

Discussion

SS is defined as a malignant mesenchymal neoplasm with partial epithelial differentiation which occurs predominantly in older children and young adults. It occurs predominantly in extremities.[1-3] The name SS is a histological error as it neither arise from nor differentiate toward synovium.[4,5] Head and neck region is the most commonly affected region after extremities, representing 5% of all cases.[6] SS is characterized by the presence of the t (X;18) (p11.2; q11.2) translocation,

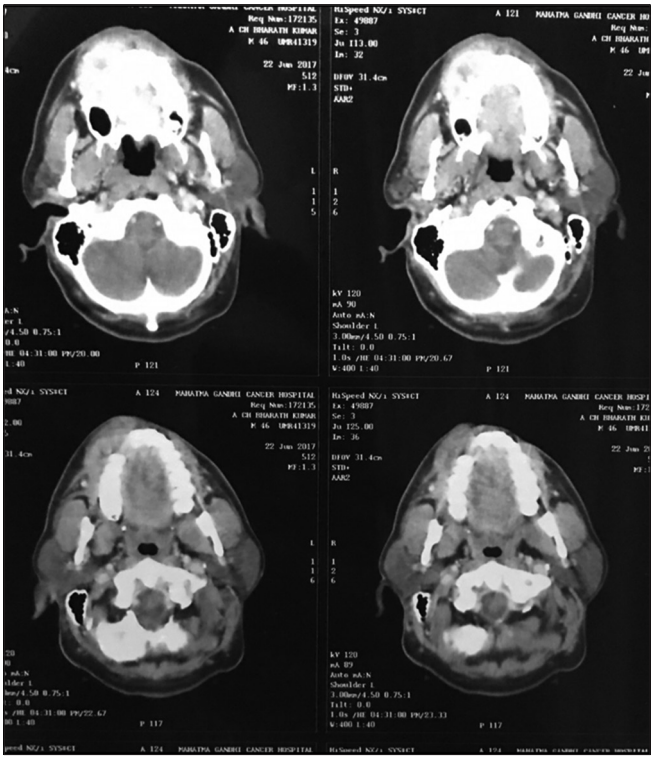

Figure 2: Computed tomography scan of paranasal sinuses

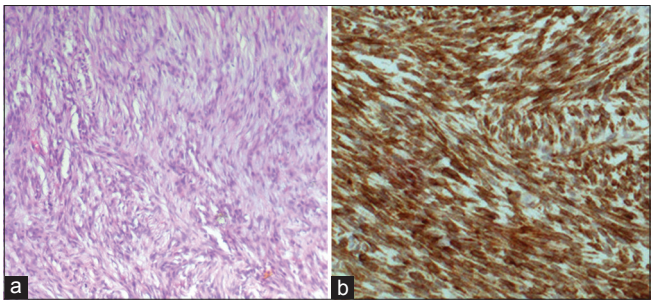

Figure 4: (a) The hematoxylin and eosin stained section presents spindle cell proliferation and small round to oval cells. (b) Spindle cells reveal Immunohistochemical positivity with Bcl2

involving the SS18 (formerly SYT) gene on chromosome 18 and one of several SSX genes on chromosome X (usually SSX1 or SSX2), which is seen in more than 90% of SS and results in the formation of SS18-SSX fusion oncogenes.[7]

SS is predominant in childhood and adolescence with age range of 15–40 years.[4,8] The case presented had a slightly higher age range compared to the mean age of the cases presented so far. SS has a slight male preponderance with a male and female ratio of 1.2:1.[9,10] The most common head and neck site is the paravertebral region, with a presentation in the pharynx (including hypopharynx, parapharyngeal, and retropharyngeal spaces);[7] other sites include the parotid, temporal region, tonsil, and cheek.[6] The case presented involved right gingivobuccal sulcus and nasolabial fold extending into maxillary sinus. Radiologically, SS tend to be large, relatively well-defined and lobulated, and most frequently located in the extremities, with epicenters close to joints.[11] Approximately 30% of patients have detectable radiologic calcifications, which may be focal or dispersed throughout the tumor, often with a fine, stippled, or opaque appearance.[12] There is a large histologic spectrum, with SS showing significant morphologic overlap with a variety of neoplasms, from small round cell tumors to spindle cell sarcomas and carcinoma.[6]

Two major subtypes of SS are observed, namely, biphasic and monophasic spindle cell types. Other rare varieties observed are monophasic epithelial, poorly differentiated (round cell), calcifying/ossifying, and myxoid types. Monophasic spindle cell type is the commonest form which consists of hypercellular arrays of relatively small spindle cells with uniform, ovoid, short, and vesiculated nuclei. Biphasic SS comprises a mixture of fibroblast-like spindle cells similar in appearance to those of the monophasic spindle cell subtype and epithelial cells, the latter often forming gland-like structures.[12,13]

The differential diagnosis includes leiomyosarcoma and rhabdomyosarcoma. It can be differentiated from muscle tumors by the presence of intersecting fascicles of cells, rather than the longer fascicles seen in monophasic SS. MPNST can be difficult to distinguish from SS. MPNST can be differentiated by S-100 protein expression which is usually focal and scanty and presence of focal CD34 positivity. These could be differentiated by immunohistochemistry (IHC). IHC was performed and revealed that the tumor cells were positive for cytokeratin and Bcl2 and negative for myoglobin, SMA, S100, CD 31, CD 56, HMB45, VEGF, and P63. A final diagnosis of "Poorly differentiated SS of rhabdoid variety" was made accordingly. The treatment of SS is multimodal which involves surgery, radiotherapy, and chemotherapy. It has tendency for late recurrence and metastasis.[13,14] The prognosis is poorest in cases treated with inadequate margins and without any adjunctive therapy, with recurrence rate 80% reported. With adequate surgical excision or with adjunctive radiotherapy, the recurrence rate has been reported to be significantly lower (<40%).[1] Therefore, long-term follow-up of more than 10 years is mandatory.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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