Synovial sarcoma of unusual site treated with chemo-radiation

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Introduction

Synovial sarcomas are high-grade neoplasms arising from primitive undifferentiated pleuripotent mesenchymal cells unrelated to synovial membranes [1,2]. Synovial sarcomas usually arise in close association with tendons, tendon sheaths, bursae and juxta-articular membranes with only 10% actually involving the joints. They are usually confined to hip, knee, ankle and shoulder joint, but about 3–10% of them are seen in Head and Neck region. In Head and Neck region, hypopharynx and parapharyngeal space are the most commonly affected area, though individual cases have been reported involving anterior neck, larynx, tongue, nasopharynx, gingival sulcus, middle ear, pyriform fossa, cheek, parotid and infratemporal area. The diagnosis is made from distinctive histopathological appearance. Histochemical, immunohistochemistry, electron microscopy and cytogenetic analysis can also be used as adjunct to the diagnosis.

Case report

A 20-year-old male presented to the outpatient department with the complaints of painless swelling in right side of oral cavity since five months, gradually increasing in size.

Swelling was also noticed in the neck since last two months. Patient complained of difficulty in swallowing due to the mass in oral cavity. On examination there was a firm soft tissue mass in the right side floor of mouth, which was extending posteriorly till the anterior tonsillar pillar. Mass was pushing the tongue upwards and backwards obstructing the airway. There was swelling in the right submandibular and submental region also. Routine haematological investigations were normal. Differential diagnosis of deep mycosis, squamous cell carcinoma or mesenchymal tumour was considered. The computed tomography scan from base of skull to thoracic inlet revealed a homogenous non-enhancing soft tissue mass in the floor of mouth right side, extending to right submandibular region and parapharyngeal space [Fig. 1]. There was no bony destruction. Computed tomography scan of chest found no metastasis.

Biopsy was taken from the oral cavity and sent for histopathological examination. Routine hematoxylin and eosin preparation revealed mucosal covering of stratified squamous epithelium. Below it was the unencapsulated tumour mass showing biphasic pattern with compactly arranged fascicles of hyperchromatic spindle cells in matrix and epithelial cells forming the gland like configuration. Numerous mast cells were seen within tumour mass with no foci of calcification. Patient was diagnosed to have synovial sarcoma of floor of mouth extending to parapharyngeal space. Due to the extensive involvement of mass patient was subjected to chemotherapy. Patient received five cycles of chemotherapy with Injection Doxorubicin (30mg/m²), Injection Decarbazine (400mg/m²) and Injection Ifosfamide (2gm/m²). Patient had severe oedema of the tumour mass for which he was tracheostomized. Patient was given radiotherapy to head and neck region. Total of 66Gy of radiation was given to the patient in 6 weeks duration. The size of the tumour mass decreased gradually. The computed tomography of patient four months after the therapy revealed no mass in the neck [Fig. 2]. Tracheostomy site was closed later as oedema subsided. Ten months post-

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treatment, the patient is under regular follow up with no recurrence till date [Fig. 3].

Fig. 1 Computed Tomography of the patient showing the mass in floor of mouth and parapharyngeal region

Fig. 2 Computed Tomography of patient after treatment showing no mass

Fig. 3 Photograph of patient after treatment with normal floor of mouth

Discussion

Synovial sarcomas comprise of about 8-10% of all sarcomas and are believed to arise from synovial differentiation of mesenchymal tissue [3]. Healthy young adults of age group <40 years are mainly affected with slight male preponderance. They usually originate in juxtaarticular structures with only 10% actually involving the joints. Areas such as head and neck, pelvis, abdominal wall are also involved, which are usually devoid of synovial tissue. Fewer than 150 cases of synovial sarcoma have been reported in head and neck region. In oral cavity, base of tongue is the most common site involved, which is followed by cheek, soft palate, retromolar area, floor of mouth and gingivobuccal sulcus [2]. To the best of our knowledge only two cases with floor of mouth involvement have been reported out of 31 cases with oral presentation [2,10].

The presentation and pathology of synovial sarcoma involving head and neck region is same as that of the other regions. Patient presents mainly with a painless mass progressively increasing in size. According to the site of involvement, patient may complain of dyspnoea and hoarseness in laryngeal, dysphagia in oropharyngeal or hypopharyngeal involvement. The mass is rubbery, spherical and well circumscribed. Histologically it exists in two forms- monophasic and biphasic patterns [4]. The biphasic form contains both sarcomatous and epithelial components whereas uniphasic form has only one of the two components. Of the two forms, biphasic pattern is most commonly seen. The epithelial cells-cuboidal or columnar are seen arranged in glandular formations, compact nests or cleft like spaces. They may be seen forming papillary projections [5,9]. The stroma is typically formed by sarcomatous fibroblasts like spindle cells. These cells formed solid compact sheets showing variable amount of mitosis. Hyalinized collagen may be seen in stroma dividing the spindle cells into fascicles. Areas of necrosis and haemorrhage may or may not be seen [4]. Psammomatous or dystrophic form of calcification is seen in about 30% of synovial sarcomas but extensive calcification is unusual.

Along with the typical histopathological appearance of the tumour, the histochemical, immunohistochemistry, electron microscopy and cytogenetic analysis are also used to confirm the diagnosis. A consistent specific balanced reciprocal translocation $t(X; 18)$ is found in more than 90% of all synovial sarcoma types occurring at all anatomical sites. It is reported to help in diagnosis, confirming tumour-free margins during surgical resection and to verify presence of metastasis disease [6].

Although the synovial sarcomas are slow growing tumours, the long-term survival rates are not reported to be good [5,7]. They have a tendency to recur locoregionally in about 20-30% of cases. Long-term survival rate falls due to distant metastasis, lungs being the most common site followed by bone marrow. There are papers published

in literature reporting these tumours to be less aggressive with better prognosis as compared to other sites [8]. It is probably due to early detection and easy accessibility of these tumours in head and neck region. Pae S et al in 1993 gave certain criteria for the favorable outcome of these tumours- size less than 5cm, extensive calcification, adequate surgical resection, absence of extensive necrosis and high mitotic index [11]. In 1987, Moore DM reported these tumours to have similar aggressive nature with poor prognosis as those involving other sites [12].

Surgical resection of the tumour is reported to be the mainstay of treatment [2,3]. Several authors recommend wide surgical resection but postoperative radiotherapy decreases the local recurrence [13]. A total dose of 65Gy has been recommended for the local control of this tumour. Problem encountered in tumours of oral cavity and neck is the large area involved which makes surgical excision difficult and sometimes impossible. In our case also, keeping in view the extensive morbidity of surgery, chemoradiation was the treatment resorted to. There are reports in literature, which support the chemotherapy as successful mode of treatment [14,15]. Besides having some radiation mucositis the patient is doing fine after ten months of treatment. Patient is under constant follow up to rule out any local recurrence or distant metastasis.

Summary

Synovial sarcoma is a rare tumour to present in Head and Neck region. Generally it presents as a painless neck mass. Biopsy of the specimen gives the way to correct diagnosis. It is rapidly growing tumour with metastasis to lymphatics, lungs and bone marrow being most common. The treatment of this tumour mainly comprises of wide surgical excision with neoadjuvant chemoradiation. Radiotherapy and chemotherapy are being used separately also for the treatment. Survival rate of these patients is considered to be good if early diagnosis and treatment is given. Long term follow up is required to detect any local recurrence and distant metastasis.

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