

Undifferentiated Pleomorphic Sarcoma of Submandibular Gland- A Rare Case Report

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Abstract

Undifferentiated pleomorphic sarcoma, previously known as malignant fibrous histiocytoma, was one of the most common variant of soft-tissue sarcoma in adults (40 %). It was first recognized as a distinct clinicopathologic entity in the early 1960s as pleomorphic sarcoma. In the head and neck, malignant fibrous histiocytoma is extremely rare and accounts for only 3 % of all undifferentiated pleomorphic sarcomas. We are reporting a large unusual case of undifferentiated pleomorphic sarcoma involving the right submandibular gland in a 60-year-old male patient.

Introduction

Sarcomas are malignant tumors of mesenchymal tissue origin, occurring at any site of the body, rarely reported in salivary glands. According to Siefert and Oehne's review of 167 mesenchymal tumors of the salivary glands, only 17 were sarcomas (10 %) [1]. World Health Organization (WHO) considers the existence of undifferentiated type of pleomorphic sarcomas and defined it as a group of high-grade pleomorphic sarcomas that should be considered as a differential diagnosis [2,3]. The predisposing factors are chronic irritation, exposure to radiation, chemotherapy, genetics, chemical carcinogens etc. Sarcomas are thought to origin as a de novo and not from a pre-existing benign tumor according to most of the cases reported in the literature so far [4]. In the past, most cases of Undifferentiated High-Grade Pleomorphic Sarcomas (UPSs) were diagnosed as pleomorphic variant of malignant fibrous histiocytomas [5]. O'Brien and Stout (1960) first described the malignant fibrous histiocytoma, and WHO named it as Undifferentiated High-Grade Pleomorphic Sarcomas (UPSs) [6]. UPSs represent about 5 % of all soft-tissue sarcomas in adults and occurs mostly in the lower extremities among patients in the sixth and seventh decades of life. In the head and neck, it was extremely rare [3] and few literature reports occurrence in parotid gland. Here, we are presenting a rare case of UPS of the right submandibular gland extending to parotid region.

Case report

A 60 years old male patient presenting with a 6-month history of a slowly enlarging painless right-sided neck mass of size 10 x 12 cm reported to our Department. Contrast enhanced CT scan of head



Figure 1: Preoperative.

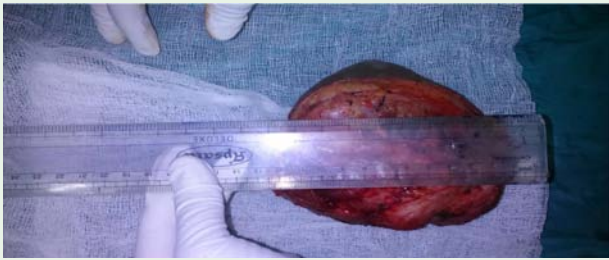


Figure 2: Intra operative.

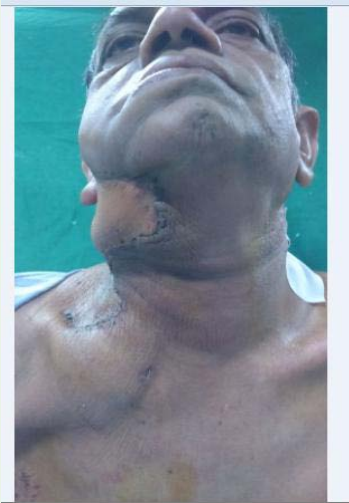


Figure 3: Post operative.

and neck showed a solid, lobulated, nodular, non-tender mass having multiple areas of focal calcifications arises from the submandibular gland having dimensions of 12.4 * 12.8 * 10.5 cm. The lesion was a large, fairly well defined, and heterogeneously enhanced, soft-tissue mass on the right side of the face and neck, with the epicenter situated in the submandibular space extending to right parotid region. These findings were suggestive of a neoplastic mesenchymal mass (Figure 1).

Surgical procedure done was wide local excision with radical neck dissection of right side and reconstruction of defect with pectoralis major myocutaneous flap under general anesthesia (Figure 2). This sarcoma of pleomorphic variant involving the submandibular gland was one of the rarest cases reported in the literature till now (Figure 3).

Discussion

Pleomorphic sarcoma mostly occurs in the extremities, trunk, and retro peritoneum, but primary lesions of the viscera, trachea, and genitals have also been reported [7]. Only 3 % involvement in head and neck region is seen with variable site specific prevalence rate. 30 % in the nasal cavity and paranasal sinuses, 15-25 % in the craniofacial bones, 10-15 % each in the larynx and soft tissues of the neck, and 5-15 % in the oral cavity [8]. Auclair et al. studied 67 cases of sarcomas of the major salivary glands and found that in 88 % cases, parotid glands are involved and 12 % cases in the submandibular glands. No cases involved the sublingual gland. Left side is more

commonly involved than right side in parotid gland sarcomas [9]. Various predisposing etiological factors have been proposed like secondarily following radiation therapy and traumatic burn injuries, associated with Paget's disease and fibrous dysplasia [10]. UPSs have five histological variants: storiform pleomorphic; myxoid; giant cell; inflammatory and angiomatoid. The storiform pleomorphic subtype of malignant fibrous histiocytoma is the most common subtype and mostly seen as deeply seated tumor of the extremities in middle-aged or elderly patients [11].

Currently, the treatment of choice for the primary site is aggressive surgical resection with radical neck dissection for cervical lymphadenopathy for large sized tumors [12]. Due to its high recurrence rate, postoperative radiation therapy is recommended to improve local control [12]. The reported rate of metastasis is as high as 44 % and commonly involves the lung, regional lymph nodes, liver, and bone [13,5]. The use of adjuvant chemotherapy and/or radiation therapy as a primary treatment modality has been less successful due to its chemo resistant nature but recent advances in these treatments are still under investigation [13]. Although UPSs are rare, the most appropriate treatment appears to be surgery associated with adjuvant radiotherapy, given that the risk of disease progression was reported to increase when adjuvant radiotherapy was not used [14]. Considering its fast growth pattern, close follow-up is essential [15]. In our patient close follow-up shows no recurrence till now.

In conclusion, there is significant controversy regarding the existence and management of MFH. Nevertheless, our case represents one of the rare reports of undifferentiated high-grade pleomorphic sarcoma involving the submandibular gland as per the literature reviewed till now.

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