Pakistan Journal of Otolaryngology 2014; 30 : 90-92

Case Report

A Giant Submandibular Gland Carcinoma Ex-Pleomorphic Adenoma

Ejaz Ahmed, Anjum Khawar

ABSTRACT: Pleomorphic adenoma(PA) is the most common benign tumor of salivary glands. It has malignant potential if recurrent or left untreated. There are three subtypes of malignant PA: carcinoma ex-pleomorphic adenoma, true malignant mixed tumor and metastasizing pleomorphic adenoma. We report a case of large carcinoma ex-pleomorphic adenoma arising from the right submandibular salivary gland in a 70 years old male. The lesion measured~32×38×29 cm (AP×T×CC) and weighed 7.6 kg. This case report emphasizes that if pleomorphic adenoma left untreated, it can transform into carcinoma ex pleomorphic adenoma. To the best of our knowledge this case of carcinoma ex pleomorphic adenoma of submandibular gland is one of the largest ever reported regionally.

KEY WORDS : Salivary gland tumors, Pleomorphic adenoma, Carcinoma ex pleomorphic adenoma.

INTRODUCTION : Salivary gland tumors are rare and make up to 3% of head and neck tumors[1]. The most common salivary gland tumor is pleomorphic adenoma (PA). This is also referred to as “benign mixed tumors.” They represent 60 to 80% of all benign tumors in major salivary glands and 40 to 70% of minor salivary gland tumors[2]. These tumors are painless and slow-growing. Failure to seek early treatment can result in gross facial deformity. They are also reported to occur in the main bronchus, larynx, pharynx, trachea, lacrimal gland, nasal tract and maxillary sinus, although less frequently. Lymphadenopathy and nerve compression are generally less prevalent Pleomorphic adenoma may become malignant if left untreated. Carcinoma ex-pleomorphic adenoma (CXPA) constitutes 3.6-4% of all salivary gland tumors and 12% of all malignant salivary gland tumors[3,4]. The exact etiology associated with malignant transformation is unclear; however exposure to radiation is thought to be a factor. We report a case of large carcinoma ex-pleomorphic adenoma arising from right submandibutor salivary gland in a 70 years old man.

CASE REPORT : A 70-years-old male presented to our Otorhinolaryngology-Head and Neck Surgery department seeking medical attention for a large swelling on the right side of his jaw (Fig-1). The swelling was painless gradually increased in size over 6 years. He noticed the swelling while shaving; he visited a local hospital where an incisional biopsy was done. Histology report showed net of epithelial and myoepithelial cells, numerous ducts with no evidence of malignancy. Size noted then was 6×4 ×2.5 cm. He ignored his swelling and did not get any treatment. In 2012, he presented to CMH hospital Rawalpindi where a FNAC was performed which showed hyperchromic and pleomorphic nuclei with deranged N/C ratio. Size noted was 11.3×9.8×9.6 cm. In November 2013 he presented in our OPD with these reports. The patient was otherwise healthy with no significant past medical history.

On examination, the weight of the swelling made the patient's head tilt to the right. The swelling was multinodular, non-tender, hard and mobile with a size of 32×38×29 cm (AP×T×CC). Movement of the tumor mass could be elicited and palpated bimanually both intraorally (in the region of the right submandibular salivary gland) and extraorally. Cervical lymph nodes were not palpable. There was no neuronal or functional deficit in the contiguous areas.

FNAC was performed; results showed pleomorphic cells with hyperchromatic nuclei and scanty cytoplasm with highly atypical cells. Axial and coronal views of the computed tomography (CT) scan revealed a well-defined heterogeneous soft tissue density mass in right side of neck anterosuperiorly extending into right submandibular region medially. Caudally this mass was abutting the adjusting surface of tongue however the fat planes of tongue were intact. Underlying bone appeared unremarkable. The mass was causing compression on the right masseter, sternocleidomastoid and strap muscles. It was adjacent to the carotid and jugular vessels however no filling defect was seen (Fig-2). A provisional diagnosis of carcinoma ex-pleomorphic adenoma was made and the patient was prepared for explorative and excisional biopsy under general anesthesia. The mass was excised and sent for histopathological examination. Weight of the excised specimen was 7.6 kg (Fig-3). Primary closure was done in a layer-wise manner with drain attached which was later removed. The histological examination was confirmatory for carcinoma ex-pleomorphic adenoma.

DISCUSSION : Pleomorphic adenoma (PA) is one of the most common benign tumors of the salivary gland. PA usually grows slowly and is painless. There are three subtypes of malignant PA: carcinoma ex-pleomorphic adenoma (CXPA); carcinosarcoma (true malignant mixed tumor) and metastasizing

Department of Otorhinolaryngology, Pakistan Institute of Medical Sciences, Islamabad - Pakistan.

pleomorphic adenoma. The most common subtype of malignant PA is CXPA which develops in primary or recurrent PA. PA can transform into CXPA over time as it enlarges. Rate of occurrence seems to increase in the period during which PA is left untreated. Furthermore, multiple recurrences may play a role in the malignant transformation of the PA[5]. CXPA is most frequently seen in the parotid gland (67%); the submandibular gland is involved in 15% of the cases. The sublingual gland is involved in only 1% of cases. Malignant transformation of PA occurs in 5 to 25% untreated patients, generally after 15-20 years and warning symptoms are present in majority of the cases[3]. FNAC has inadequate sensitivity in detecting malignant nature of PA. Clinical picture and histopathology establish the diagnosis of CXPA. A case report discusses a large CXPA that transformed from PA developed in 17 years[6]. In this case PA transformed into CXPA over a period of 5 years. The deformity caused social withdrawal and fear of surgery made the patient decline treatment. CXPA is a rare, aggressive, poorly understood malignancy. In CXPA, an epithelial malignancy develops in association with a primary or recurrent benign pleomorphic adenoma. CXPA is very difficult to identify before surgery because the clinical presentation mimics that of pleomorphic adenomas. Generally malignant transformation can be suspected with a sudden increase in size accompanied by local signs of malignancy such as pain, ulceration, spontaneous bleeding and superficial or deep tissue invasion. The clinical characteristics of malignant transformation have been reported in the literature as[1] a long history of PA[2], advanced age[3], location in a major salivary gland[4] and history of rapid growth associated with pain or ulceration[7]. In one of the major studies involving 6982 primary salivary gland tumors in a Chinese population PA was the most common tumor (69%) of which only 20% were located in minor salivary glands[8]. They have been reported to be as large as 28×20×16 cm in the parotid gland[9]. Gupta et al reported weights of PAs ranging from 1 kg to 27 kg[10]. The occurrence of very large PAs in the submandibular gland are rare and reports of 8×6×4 cm have been documented[10]. In our case site of CXPA is unusual i.e. submandibular gland with change from PA to CXPA in 5 year. It measured 32×38×29 cm (AP×T×CC) and weighed 7.6 kg. Age in our case patient was 65 years at time of first presentation but at the time of surgery it was 70 years. Tumor resection is the solitary option in CXPA. Facial palsy, salivary fistula, and Frey syndrome have been reported as complications of tumor resection. Reporting a case of this nature has significance as delayed intervention allows for rapid tumor growth, poor aesthetics, psychological trauma and a possible threat to life especially with change into malignant tumor. The expanding mass could further distort the anatomy thus displacing vital structures and making the dissection more challenging. The best method of prevention and treatment of CXPA is early detection and radical removal of all major salivary glands tumor.

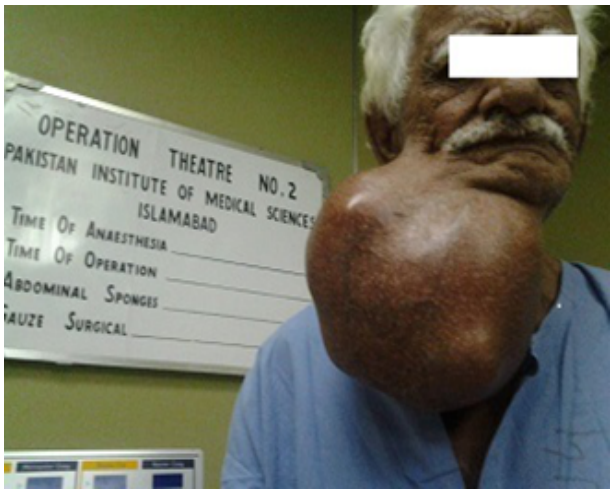

Figure-1: Clinical presentation, front and profile view of tumor mass.

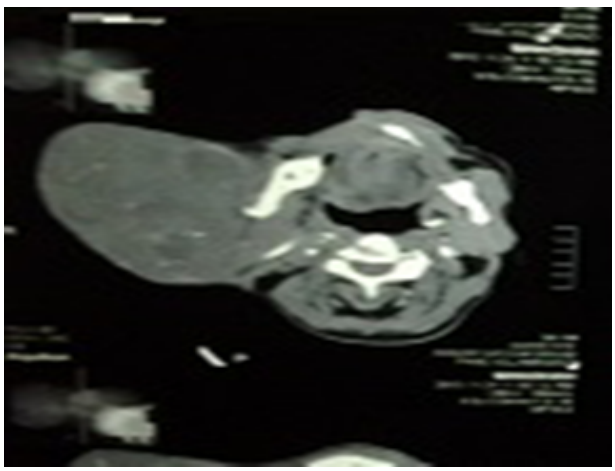

Figure-2: Axial CT scan.

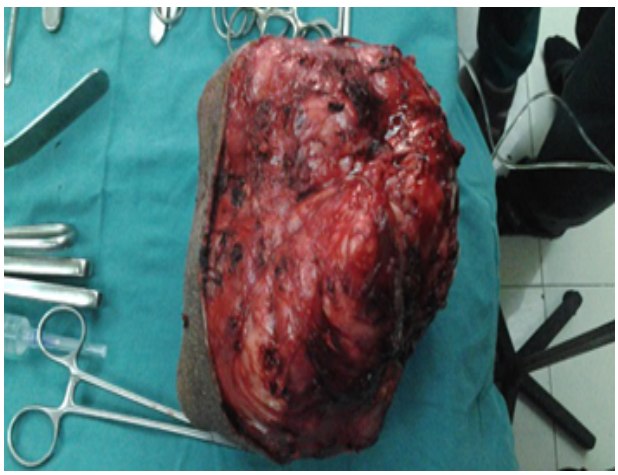

Figure-3: Excised tumor.

REFERENCES:

1. Eveson JW, Cawson RA. Salivary gland tumors: A review of 2410 cases with particular reference to histological types, age and sex distribution. J Pathol. 1985;146:51–8.
2. Molina E J, Mayer K, Khurana J, Grewal H. Pleomorphic adenoma of the submandibular gland. J Pediatr Surg. 2008;43:1224-6.
3. Stodulski D, Rzepko R, Kowalska B, Stankiewicz C. Carcinoma ex-pleomorphic adenoma of major salivary glands-a clinicopathologic review. Otolaryngol Pol. 2007;61:687-93
4. Chaudhry AP, Vickers RA, Gorlin RJ. Intraoral minor salivary gland tumors. An analysis of 1,414 cases. Oral Surg Oral Med Oral Pathol. 1961;14:1194-226.
5. Spiro RH, Huvos AG, Strong EW. Malignant mixed tumor of salivary origin: a clinicopathologic study of 146 cases. Cancer. 1977;39:388-96.
6. Yoshida N, Hara M, Kanazawa H, Iino Y . Large Carcinoma ex-Pleomorphic Adenoma of the Parotid Gland: A Case Report and Review of Literature. J Oral Maxillofac Surg. 2013;71:2196.
7. Ethunandan M, Witton R, Hoffman G, Spedding A, Brennan P A. Atypical features in pleomorphic adenoma—a clinicopathologic study and implications for management. Int J Oral Maxillofac Surg. 2006;35:608-12.
8. Tian Z, Li L, Wang L, Hu Y, Li J. Salivary gland neoplasms in oral and maxillofacial regions: a 23-year retrospective study of 6982 cases in an eastern Chinese population. Int J Oral Maxillofac Surg. 2010;39:235-42.
9. Takahama A, da Cruz Perez D E, Magrin J, de Almeida O P, Kowalski L P. Giant pleomorphic adenoma of the parotid gland. Med Oral Patol Oral Cir Bucal. 2008;13:58–60.
10. Gupta M, Chaudhary N. Giant presentation of pleomorphic adenoma in major salivary gland. BMJ Case Reports. 2011