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Original Article

Giant Pleomorphic Adenoma of Major Salivary Glands: A Review of Ten Cases

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Abstract

Background: Pleomorphic adenoma (PA) of the major salivary glands has been documented to occur in various sizes ranging from 1 cm to 10 cm in their widest diameter; however, they can assume a grotesque proportion as a consequence of uninhibited growth in cases with delayed presentation. **Patients and Methods:** A retrospective study of all patients with PA of the major salivary glands with sizes >10 cm in its widest dimension and seen over a 10-year period was undertaken by retrieving sociodemographic and clinical data from patients' case files from the Oral and Maxillofacial Unit of a Nigerian Teaching Hospital. **Results:** Case files of ten patients (4 males and 6 females) were reviewed over the study period. The ages ranged from 14 to 61 years, mean 33.8 ± 17.97 years. Equal distribution of five cases of PA was noted in the parotid and submandibular glands, respectively. The onset of symptoms ranged from 3 to 20 years and the largest tumor was 27 cm in its largest diameter. Only one case of malignant PA was recorded. **Conclusion:** Prevalence of giant PA from the study was found to be 13.5%, with a greater occurrence in the female (60%) patients compared to the male (40%) patients and with an equal distribution in both parotid and submandibular glands. PA of major salivary glands can assume a grotesque proportion when timely surgical intervention is not sought. Complete surgical excision in benign lesions can guarantee a cure and restore self-esteem.

Keywords: *Giant, pleomorphic adenoma, salivary gland*

Introduction

Pleomorphic adenoma (PA) is an epithelial tumor of complex morphology comprising of epithelial and myoepithelial elements intermingled with mucoid, myxoid, or chondroid tissue arranged in a variety of patterns within a mucopolysaccharide stroma.[1] PA is the most common benign tumor of salivary gland origin accounting for 90% of all salivary gland tumors[2] and 60–70% of tumors involving major salivary glands. The parotid gland is the most common site followed by the submandibular gland.[3] In the parotid gland, PA mostly occurs in the lower pole of the superficial lobe of the gland, and only about 10% occur in the deep lobe.[4] They are known to be rare in the sublingual gland but frequently found in minor salivary glands of the palate, buccal mucosa, and upper lip.[4] The palate is the most common site for the minor salivary glands (60–65%)[4] They are reported to also occur in the main bronchus, columella, larynx, pharynx, trachea, lacrimal gland, sinonasal tract, and maxillary sinus, albeit less commonly.[5]

Gender distribution of PA is approximately 6:4 with the lesion occurring more in females.[6] The majority of the lesion are found in patients within the third to sixth decades.[6] Patients often give a history of a small, painless, quiescent nodule, which slowly increased in size. When in the major salivary gland, it typically does not show fixation to either the deeper tissues or

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Access this article online

Quick Response Code:

Website: www.ajmhs.org

DOI: 10.4103/2384-5589.197967

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How to cite this article: Omeje KU, Efunkoya AA, Amole OI, Osunde OD, Akhiwu IB, Agbara RC. Giant pleomorphic adenoma of major salivary glands: A review of ten cases. Afr J Med Health Sci 2016;15:92-6.

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the overlying skin. The clinical appearance is often that of an irregular nodular lesion, which is firm in consistency. Cervical lymphadenopathy and nerve compromise are generally absent although distant metastasis to the vertebral column and lungs has been documented in benign cases especially with repeated surgical trauma.[7] Cervical lymphadenopathy and nerve involvement may be seen in cases of malignant transformation. Historically, the main clinical problems with PA have been the risk of recurrence and malignant transformation.[3] The high risk of recurrence may be related to its unique features, which includes incomplete capsulation and its heterogeneous histology.[1]

PA has been documented to occur in sizes ranging from 1 cm - 10 cm but can attain a grotesque proportion and weigh several kilograms. This tumor can cause facial disfigurement and if untreated can impact negatively on patients' psychology. Diagnosis of PA of the major salivary gland is often clinical but confirmed by fine-needle aspiration cytology while management is surgical with excision of the involved gland.

This study is a retrospective review of 10 patients with giant (larger than 10 cm) PA of the major salivary glands seen in a Nigerian tertiary health care facility over a 10-year period with a view to highlighting the possible extensive growth of unoperated PAs. This study, as much as the authors are aware, constitutes the second largest single review of Giant size PA in the literature after that of Schultz-Coulon.[6] In 1989, Schultz-Coulon reviewed 31 cases of giant PAs of the parotid gland. The author found a female predominance (64.5%), with an age range from 20 to 40 years old, and a tumor weight between 1 and 27 kg.

Patients and Methods

This study was a retrospective review of case files (our institution utilizes case files for the documentation of patients' records) of all patients who presented to the Oral and Maxillofacial Surgey Clinic of the Aminu Kano Teaching Hospital, Kano over a 10-year period (from January 2005 to December 2014) with a histologic diagnosis of PA. PA is a major salivary gland lesion with macroscopic features consisting of an irregular to ovoid mass with well-defined borders and mostly incomplete fibrous capsule; also microscopic features consisting of demonstrable combinations of glandular epithelium and mesenchyme-like tissue with varying proportion of each component in individual tumors was considered diagnostic. The information retrieved from the patients' case files included the sociodemographics as well as clinical characteristics such as the anatomical site, side, presenting features, the onset of symptoms and type of treatment carried out as well as treatment outcome.PA of sizes >10 cm in its largest diameter was considered to be "giant" (considering that most literature do not report lesions >10 cm). Subjects with missing or incomplete data were excluded, also excluded were recurrent cases of PA. The data were entered into Statistical Package for Social Sciences version 15.0 (SPSS Inc., Chicago, IL, USA) and analyzed. Absolute numbers and simple percentages were used to describe categorical variables. Quantitative variables were described using measures of central tendency (mean, median) and measures of dispersion (range, standard deviation) as appropriate. No tests of significance were done.

Results

A total of 74 patients with a diagnosis of PA were seen within the period of study, of which 28 were PA from the minor salivary glands. Out of the 46 PA of the major salivary glands, only 13 were considered to be of significant size and met our inclusion criteria for the study. Three of the patients that formed the study cohorts were excluded on account of missing or incomplete information. Thus, ten cases of PA of the major salivary glands met the inclusion criteria. Prevalence of PA of the major salivary gland from the study was 62.2% while that of the giant PA of the major salivary gland was found to be 13.5%. Among those affected by giant PA of major salivary gland were females (n = 6; 60%) and males (n = 4; 40%) and their age ranges from 14 to 61 years with a mean of 35.8 ± 17.97 years. There was equal involvement of the submandibular and parotid glands with a ratio of 1:1. None of the patients presented with PA of the sublingual glands. Both the parotid and the submandibular gland lesions recorded a greater right side predominance of three subjects out of the five subjects in either category. Nine out of the ten patients had a postoperative histology of benign lesion whereas only one postoperative diagnosis of a malignant lesion in a parotid gland was made. The sizes, duration, and weight of the lesions are shown in Table 1. All except two patients with ulceration in parotid and submandibular glands presented as a painless jaw swelling. Two of the female patients were noted to have always dressed in a manner to obscure the tumor or developed minimal outdoor interactions because of their tumors. All the patients had an excisional biopsy with complete removal of the involved glands. Although total parotidectomy was done for 2 of the benign PA arising from the parotid gland, precautions were taken to ensure the preservation of the facial nerve. No recurrence was observed throughout the follow-up period. The follow-up period was between 6 months and 4 years. There was no documentation of follow-up after 4 years in the patients' records.

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Table 1: Demographic, clinical, and histologic characteristics of patients with giant pleomorphic adenoma (*n*=10)

Age	Sex	Site	Size (cm)	Duration (years)	Ulceration	Weight (kg)	Histology
33	Female	Submandibular	8×10×12	6	No	2.2	Benign
28	Male	Submandibular	15×12×13	5	No	2.9	Benign
45	Male	Parotid	12×12×14	5	No	2.4	Benign
35	Female	Submandibular	11×9 × 13	7	No	2.1	Benign
60	Male	Parotid	18×15×15	10	Yes	2.1	Malignant
61	Male	Parotid	23×22×20	18	No	3.2	Benign
45	Female	Submandibular	27×20×20	20	Yes	3.5	Benign
18	Female	Parotid	12×11×10	3	No	2.0	Benign
19	Female	Parotid	11×10×9	4	No	2.0	Benign
14	Female	Submandibular	10×12×7	4	No	2.0	Benign

Our patients were treated by surgical excision of the lesion with the involved gland while paying adequate attention to hemostasis as these tumors often have well-developed feeder vessels.

Discussion

PA is the most common benign salivary gland neoplasm accounting for 45–74% of all salivary gland tumors,[3,4] and usually presenting as a painless slow growing mass. It is frequently found in the major salivary gland[3,8] with a peak age incidence of 30–60 years and a female predominance.[3] In a review of 31 giant PAs occurring in the parotid gland over a period of 140 years by Schultz-Coulon in 1989, most occurred in females (64.5%) and only 35.5% occurred in males[6] which is consistent with the gender variation of other salivary gland tumors (except Warthin's tumor) and particularly with that of PA.[4] The age of first tumor manifestation for PA was noted by Schultz-Coulon[6] to vary between 20 and 40 years (Reference). However, pediatric patients could also be affected.[9]

The demographic characteristics of the patients in the present study concurred with the age and gender prevalence in the literature.[6] a,bThe literature also documents PA to occur in sizes up to 10 cm in its widest diameter;[3] however PA of 16 cm has been reported.[10] The largest of the tumors seen in this study presented with dimensions of 27 × 20 × 20 [Figure 1], which is larger than previously reported sizes.[3] The observation is not surprising; giving that the said case has had the tumor for about 20 years, which was longer than any of the other cases.[3] It is logical to assume that long-standing tumors may proportionally grow to large sizes.

PA though known to be firm in consistency, areas of cystic degeneration may sometimes be palpated if they are superficial. They can grow and assume an unusual dimension and weight when early intervention is not sought. When very large they appear as a single but

Figure 1: Preoperative photographs of one of the patients with left submandibular pleomorphic adenoma

irregularly nodular mass, which stretches the overlying skin or mucosa. The unsightly appearance of such a large tumor may result in an attempt by patients to often cover the lesion and when this is difficult because of considerable large sizes, this may limit patients outdoor and other social interactions. The covering of the tumor and limitation to outdoor and social interactions was noted in 2 of the female patients.

The late presentation appears to be a major factor in the size of the PA presented. Oji[11] has observed that some of the factors affecting late presentation include ignorance and poverty. Olaitan *et al*.[12] also identified low socioeconomic status, distance to the hospital, and different myths related to fear of hypodermic needles and nasogastric tube used in the hospitals as other reasons for delayed presentation in patients with orofacial tumors.

Malignant changes can occur in PA and include three distinct pathologic entities: Carcinoma arising in PA, which has been reported to occur in about 3–4%, carcinosarcoma, and benign metastasizing PA.[4,10] Malignant changes were found in only 10% (*n* = 3) of

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the giant PA cases reviewed by Schultz-Coulon.[6] The incidence of malignancy frequently shows a correlation between the length of the history of PA and the development of a carcinoma.[13] It has been reported that the risk of development of malignancy is only about 1.5% up to 5 years but increases to 9.5% after more than 15 years.[14] There was only one case of malignant PA in our series. The patient with malignant PA in our series gave a history of a sudden onset of pain after about 10 years of painless jaw swelling. The pain necessitated hospital presentation. Transformation to malignancy, in this case, may be responsible for the sudden onset of pain. Other factors that may suggest malignant transformation includes a sudden increase in size, the involvement of nerve or ulceration of the overlying skin, spontaneous bleeding, and superficial and deep tissue invasion.[10,15]

The rare occurrence of skin necrosis with ulceration and pain in two of our cases was not related to malignant transformation in one (as confirmed by histopathology) but may be due to the inability of the supporting vascular elements to cope with an increased vascular demand from the excessively large tumor. It is also thought to be as a result of pressure effect of tumor on the already stretched skin leading to further vascular compromise. The ulceration in the second case was thought to be as a result of the destruction of vascular elements in association with malignancy. The notable common causes of such ulceration in our environment are the application of topical corrosive traditional medication or heated hot irons by traditional healers. History of such was denied in our cases.

Surgical excision is the treatment of choice, often requiring concomitant excision of the involved gland. A more radical surgery is done in the case of a malignant variety.[2] Intraoperative measures to limit blood loss for such a large mass may include preoperative hemo-dilution, reversed Trendelenburg patient positioning and suspension of mass above the heart for few minutes before dissection to encourage the return of blood from the tumor back into the systemic circulation. Others measures include meticulous dissection and intraoperative hemostasis.

The size at the presentation of these tumors [Figures 1 and 2] underscores the need for strong public health drive in remote parts of Northern Nigeria. Provision of rural health centers and training of indigenous personnel for the timely detection and referral of such cases to tertiary centers should characterize this drive.

Conclusion

Prevalence of giant PA from the study was found to be 13.5%, with a greater occurrence in the female (60%) patients compared to the male (40%) patients and with an equal distribution in both parotid and submandibular glands. PA of major salivary glands can assume a grotesque proportion when timely surgical intervention is not sought. Complete surgical excision in benign lesions can guarantee cure and restore self-esteem.

Figure 2: Preoperative clinical photographs of a patient with pleomorphic adenoma of the parotid gland

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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