Carcinoma Ex pleomorphic Adenoma- A Rare malignant tumor of parotid gland-Case Report

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Abstract

In salivary gland tumors, pleomorphic adenoma is a very common benign tumor. Parotid is the most commonly involved gland by this tumor. But malignant transformation in a long standing pleomorphic adenoma is rare. The prognosis of this tumor depends on its invasiveness in the adjacent tissue. Most commonly we get poorly differentiated adenocarcinoma or undifferentiated carcinoma.[1] We report a case of 67years old male patient with a cystic swelling in the submandibular region since 20 years with sudden enlargement in the swelling since last 3 months. The histopathological diagnosis given was carcinoma ex pleomorphic adenoma.

KEYWORDS: Ca Ex Pleomorphic adenoma, parotid, salivary gland

INTRODUCTION

Carcinoma ex pleomorphic adenoma is defined as a pleomorphic adenoma from which an epithelial malignancy is derived. It comprises approx. 3.6% of all salivary gland tumors.12% of all salivary malignancies and 6.2% of all pleomorphic adenomas.Ca Ex pleomorphic adenoma usually presents in 6^{th} or 7^{th} decades.[1]

Many Ca Ex pleomorphic adenoma probably result from the accumulation of genetic instabities[1] in long standing pleomorphic adenoma.It frequently arises in the parotid gland but may also originate from submandibular and other minor salivary glands. Typical history is that of a longstanding mass present much longer than 3 years, with rapid growth over the previous few months. Patient frequently complains of a painless mass but pain, facial nerve palsy and skin fixation may also occur.[1]

CASE HISTORY

67 years old male patient came with complaints of cystic swelling at left parotid region since 20 years with sudden increase in size since last 3 months. There was no history of pain, facial palsy.

Patient was clinically diagnosed as parotid gland tumor-? pleomorphic adenoma. Hence wide local excision of the tumor was done.

Other routine investigations were normal.

On gross-It was 8x6x3 cm mass, bosselated on external surface and cut surface whitish along with few areas of hemorrhage. Adjacent to the mass normal salivary gland was noticed. No invasion was noticed in the adjacent tissue.

Microscopically-partially encapsulated tumor mass composed of squamous, intermediate, mucin producing and clear cells. At places,many squamous pearls with individual keratinisation are noted. The cells show nuclear pleomorphism, hyperchromatism with moderate amount of cytoplasm. The cells are arranged in glandular pattern with come do pattern at places, foci of hemorrhage are also seen.Periphery of lesion shows features of pleomorphic adenoma with abundant chondromyxoid matrix and normal salivary gland. There were no vascular emboli, perineural invasion and capsular invasion. So pleomorphic adenoma with foci of squamous cell carcinoma and adenocarcinoma were noted in the present case.

Final diagnosis given was Carcinoma Ex Pleomorphic Adenoma(noninvasive).

DISCUSSION

Caricinoma Ex Pleomorphic adenoma also called as malignant mixed tumor constitute approx 12%of malignant salivary gland tumors.6.2%of all mixed tumors.3.6% of all salivary gland tumors.It most frequently arises in the parotid gland. Malignant transformation occurs in 2-7% of mixed tumor. These most commonly involve parotid(73%), submandibular gland 16%and minor salivary gland 11%.[2]

Tumor may present at any age, but usually range is 7-86 years.[2]

Our case also falls in the same age group.

The most common clinical presentation of Carcinoma Ex Pleomorphic Adenoma is of firm mass in parotid gland.[6]

Nouraei et al [6]reported the presence of a parotid mass in 96%of the 25 patients examined, whereas Oslen and Lewis noted a parotid mass in 86% of 66 patients examined.[6,7.]

In our case parotid was involved.

Rarely ,patients of Carcinoma Ex Pleomorphic Adenoma may carry a slow growing mass for over 40 years before coming to clinical attention.[7] In our case also the mass was for last 20 years.

Gross-On average Carcinoma Ex Pleomorphic adenoma is more than twice the size of its benign counterpart ranging upto25 cm in greatest diameter. They are poorly well circumscribed and many are extensively infiltrative, hard ,white to tan

grey. Our tumor also showed 8x6x3cm size with grayish white areas with no evidence of invasion.

Microscopy-More reliable features of transformation into a malignant tumor are overtly invasive growth, necrosis, vascular permeation, perineural extension and prominent cytologic atypia associated with abnormal mitotic fig. Large areas of hyalinised scarlike fibrosis within a mixed tumor should lead to careful examination for a malignant component .The earliest microscopic evidence of a malignant transformation is often characterized by aggregates of large, hyperchromatic, cytologically atypical cells embedded in hyalinised stroma[3] .proportion of benign versus malignant components can be quite variable ,so extensive sampling[1] is necessary to find the benign component and in rare cases benign remanant might not be found. But if there is clinicopathologic documentation of a previously excised pleomorphic adenoma in the same site ,then the malignancy can also be classified as a Ca Ex Pleomorphic adenoma.In the present case foci of benign pleomorphic adenoma along with elements of squamous cell carcinoma and adenocarcinoma are noted.

The malignant component is most commonly a poorly diff adenocarcinoma or undifferentiated carcinoma.[3]

An infiltrative ,destructive growth pattern with nuclear hyperchromatism and pleomorphism are the most imp dignostic criteria .Ca Ex Pleomorphic adenoma is sub classified into[1]

1) noninvasive
2) minimally invasive(<1.5mm invasion of malignant counterpart into extracapsular tissue)
3) invasive(>1.5 mm of invasion from the tumor capsule into adjacent tissue)
   Prognosis depends upon pathological staging parameters like
   1. level of invasion.
   2. Lymph node invoivement.
   3. Local or distant metastasis.[1]

   prognosis is excellent in patients whose malignant focus is within tumor however it is bad when it invades the capsule.patient with high grade carcinomatous component have poor prognosis when compared to patient with low grade carcinoma.[8] Furthermore tumor size and grade are also noted to be important prognostic indicator in Carcinoma Ex Pleomorphic Adenoma.
   Our case was of noninvasive type and showed both patterns of adenocarcinoma and squamous cell carcinoma of low grade. Follow-up of the patient in last six months appears good.

CONCLUSION

Pathological assessment is the gold standard for making the diagnosis of Carcinoma Ex Pleomorphic Adenoma.

Overall the survival rate for Carcinoma Ex Pleomorphic Adenoma is worst than for most salivary gland malignancy.

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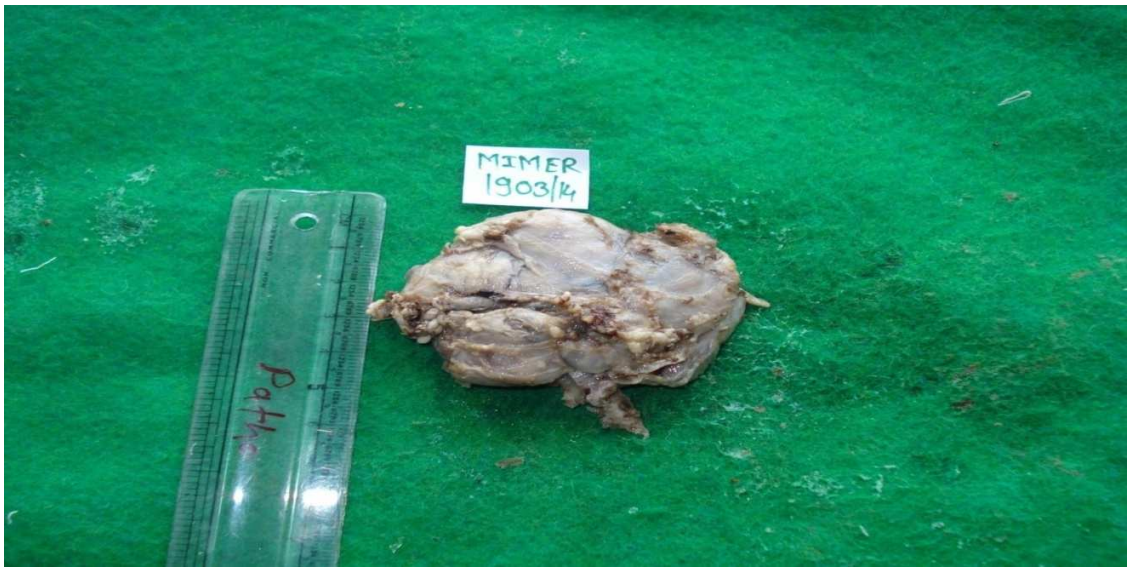
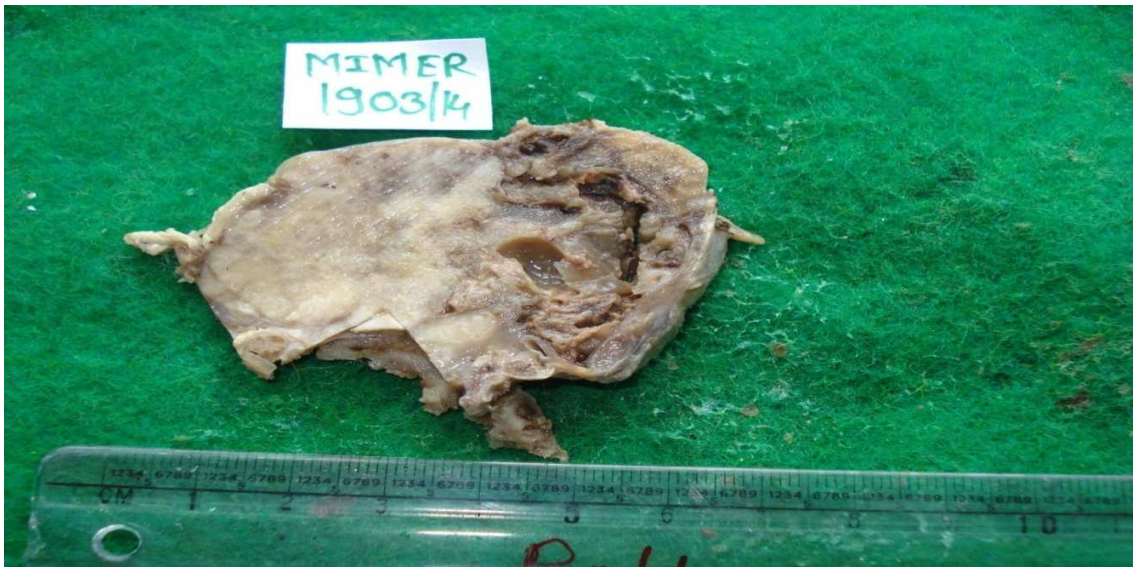
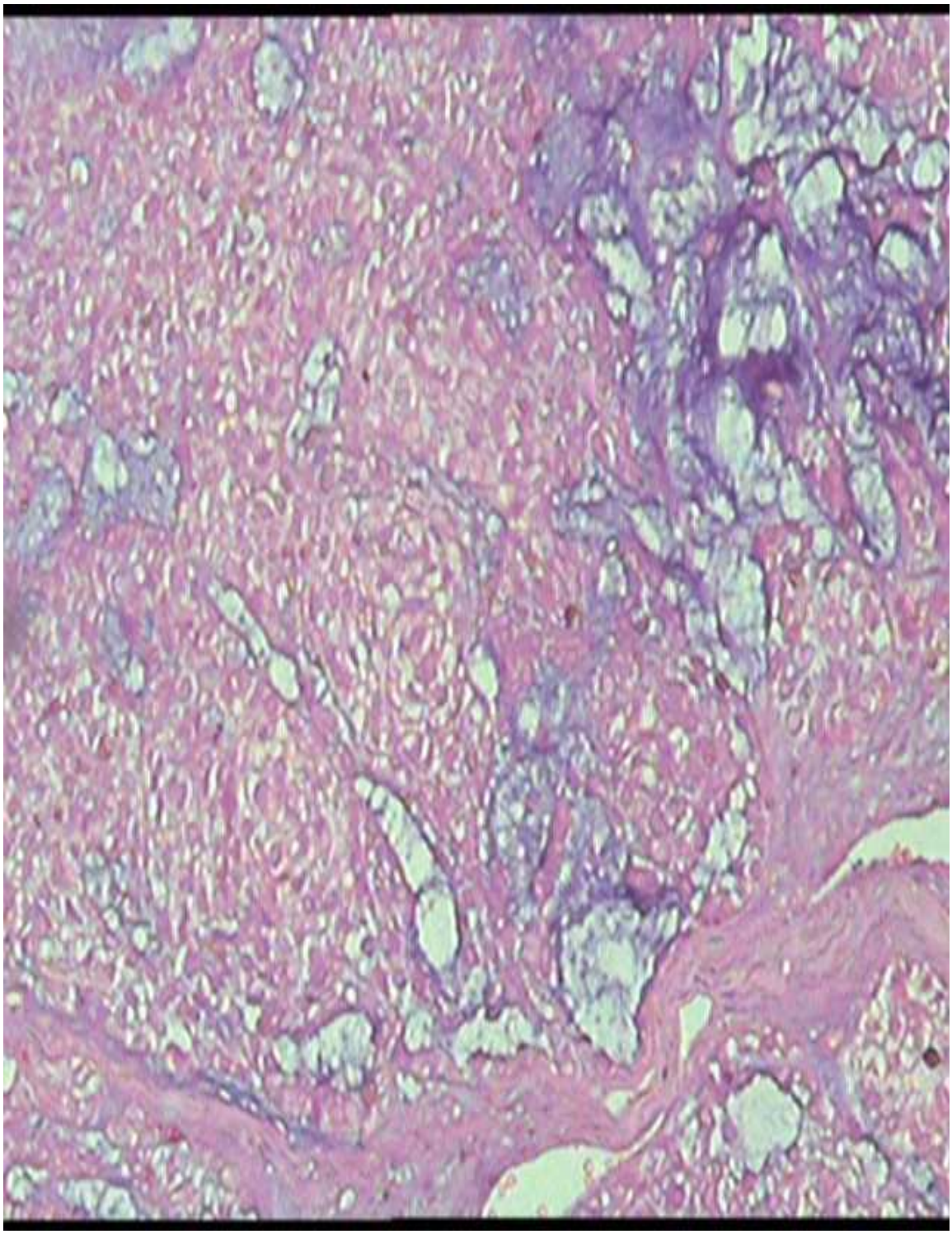
External surface is nodular, partially encapsulated.



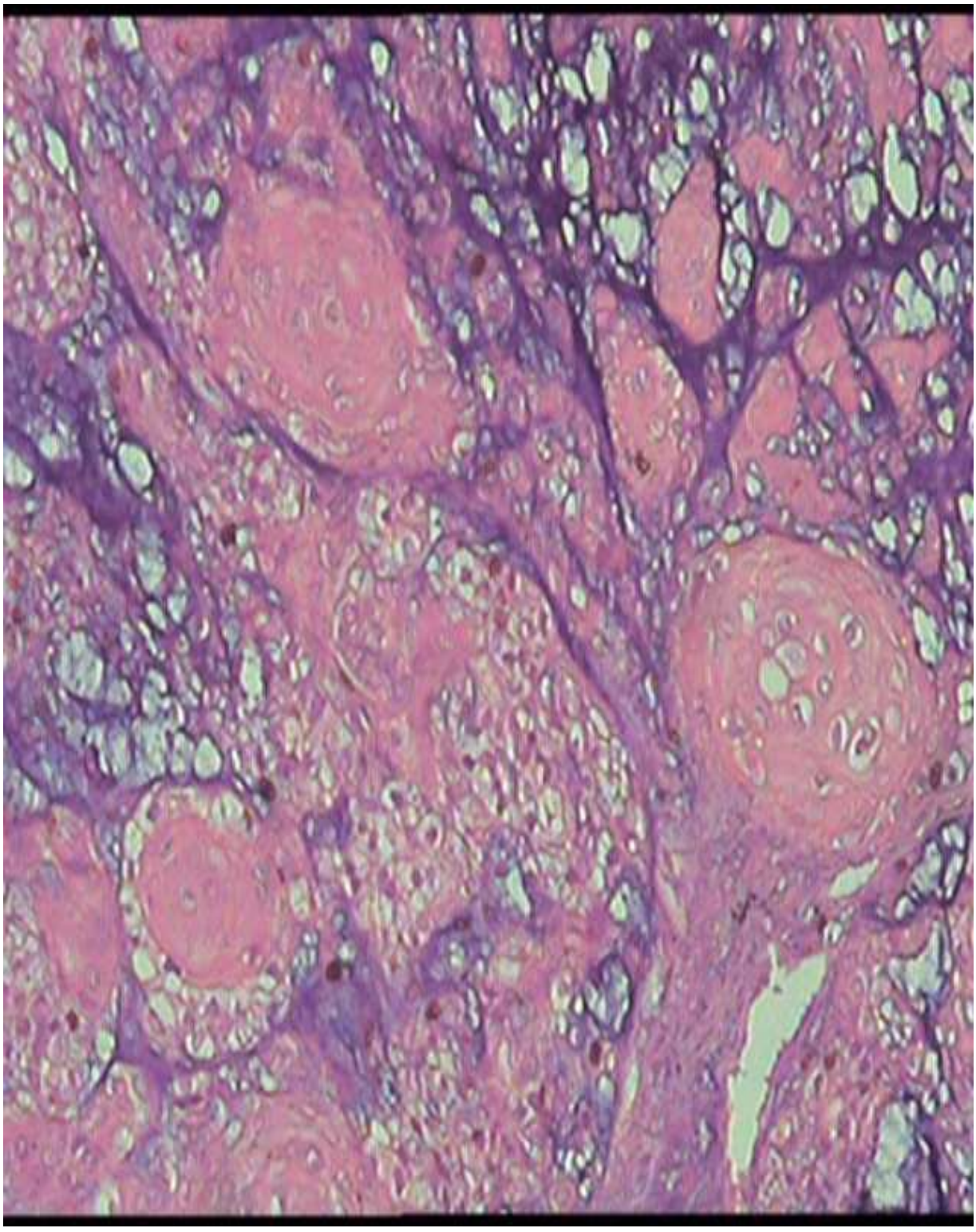
Cut surface shows grayish white tumor mass with cystic areas.



H&E (10X) Solid sheets of tumor cells with mucinous areas.



H&E 40X Squamous Pearls



H&E Stain 40x Tumor cells with clear cytoplasm.