Case Report

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Carcinoma Ex-Pleomorphic Adenoma of Parotid Gland with Hepatic Metastasis: Clinico-Radiological Case Report

Oncology Section

MANU DHILLON[1], DIVYA TOMAR[2], MANU SHARMA[3], SAMTA GOEL[4], SIDDHARTH SRIVASTAVA[5]

ABSTRACT

Pleomorphic adenoma originally called the mixed tumour is a neoplasm commonly involving major salivary glands. The spectrum of malignancy in pleomorphic adenoma comprises three distinct entities – Carcinoma ex-pleomorphic adenoma, carcinosarcoma and benign metastasising pleomorphic adenoma. Carcinoma ex-pleomorphic adenoma consists of pleomorphic adenoma with a malignant epithelial component. Occasionally, carcinomas ex-pleomorphic adenoma develops metastasis. Here we are reporting a case of benign pleomorphic adenoma arising in parotid gland which turned into malignancy after four years. The patient developed facial nerve paralysis suggesting malignant transformation. Along the course of the disease, the patient developed regional metastasis to lymph nodes and neck and distant metastasis to liver. This case report emphasises the role of advanced imaging modalities in the early diagnosis of the condition and evaluation of metastasis. The patients with this condition should be treated early for favorable outcome and investigated for distant metastasis.

Keywords: Salivary glands, Pleomorphic adenoma, Mixed tumour, Malignancy

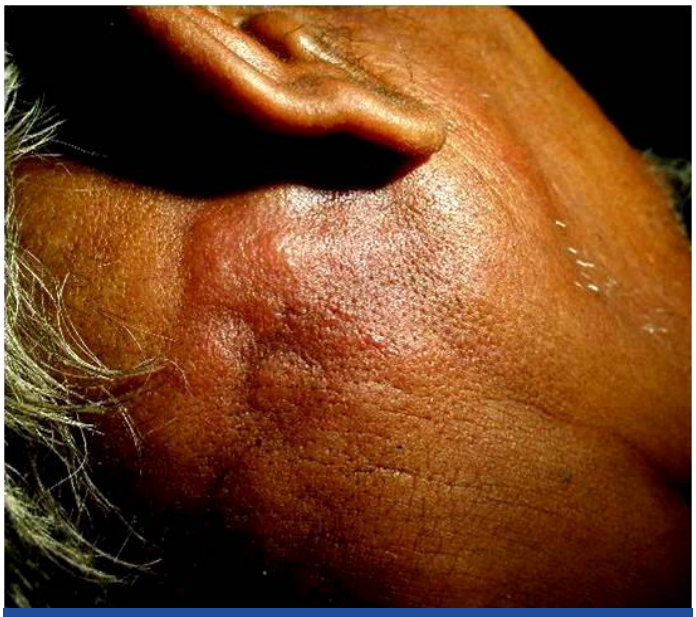
CASE REPORT

A 55-year-old male patient was presented to the Department of Oral Medicine and Radiology with a swelling on the right side of the face below the ear within last 4 years. Initial rate of increase in size was slow but since two months there was a rapid increase causing pain during tilting of head and decreased muscular tone on right side of face. On inspection, swelling was approximately 5x6 cm in size extending from preauricular region to submandibular region inferiorly and posteriorly to sterno-cleido-mastoid muscle [Table/Fig-1]. The multi-nodular swelling had irregular diffuse borders with hard consistency and was fixed to underlying structures. Right submandibular and ipsilateral superficial cervical lymph nodes showed fixity. Signs of facial nerve involvement such as loss of wrinkling and incomplete closure of the right eye were evident [Table/Fig-2]. The case was provisionally diagnosed as a case of malignant pleomorphic adenoma with metastatic lymphadenopathy and differential diagnosis of adenoid cystic carcinoma and mucoepidermoid carcinoma were given.

Investigation revealed eosinophilia [13 percent] and raised ESR (32 by Wintrobe method). Mantoux test and HIV titers were negative. No abnormal findings were evident on orthopantomogram. A Computed Tomography (CT) from base of skull showed diffusely enlarged right parotid gland involving both superficial and deeper lobes with complete loss of internal architecture [Table/Fig-3]. Invasion of right sterno-cleido-mastoid muscle and sub-mandibular gland by tumour mass was also evident [Table/Fig-4]. Extensive enlargement of submental, submandibular, cervical, internal jugular and parotid lymph nodes was present [Table/Fig-5].

Incisional biopsy of the swelling was performed and histopathological examination revealed pleomorphic adenoma in a chondromyxoid, fibrotic and sclerotic stroma with the malignant areas of carcinoma component consisting epithelial clusters with necrosis [Table/Fig-6]. The histopathological report confirmed the diagnosis of carcinoma ex-pleomorphic adenoma.

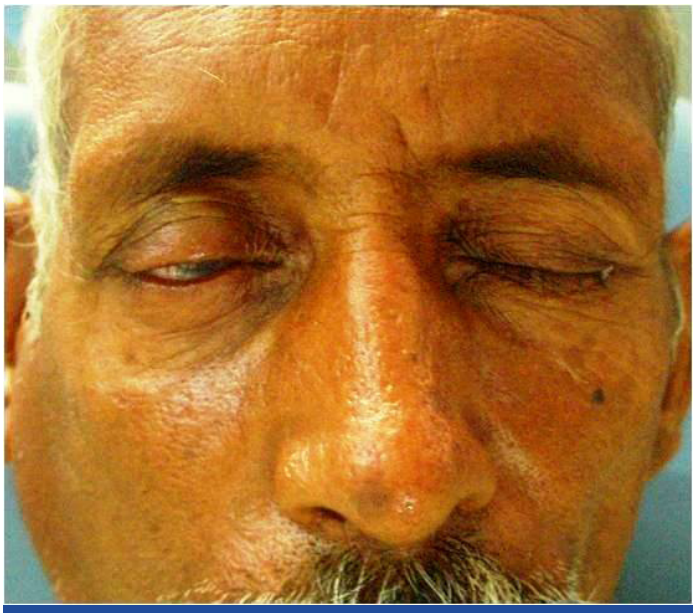
Posteroanterior chest radiograph and ultrasonography of abdomen was advised to rule out any metastatic component. Chest radiograph was clear. Ultrasonography abdomen revealed a solid, echogenic, ill defined focal lesion in postero-superior segment of right hepatic lobe in subphrenic location which was strongly suggestive of metastasis [Table/Fig-7]. FNAC of the hepatic lesion revealed features of a mixed tumour with abundant metachromatic and fibrillary stroma and small round cells as well as large pleomorphic cells in glandular pattern.



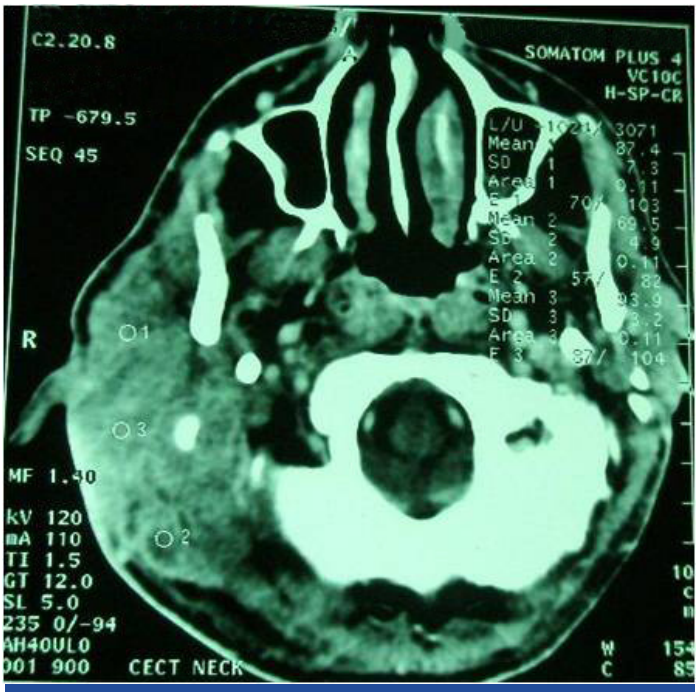
[Table/Fig-1]: Diffuse, multinodular growth extending from the preauricular region to the sternocleidomastoid muscle posteriorly

Based on the clinical, radiological and histopathological findings patient was diagnosed as a case of carcinoma ex-pleomorphic adenoma with metastatic lymphadenopathy and distant metastasis to liver.

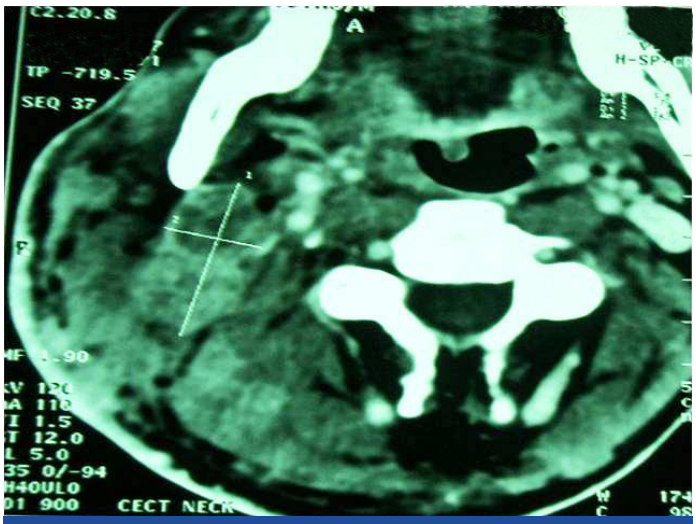
Patient was referred to specialized centre for treatment but died three months after the initiation of the treatment.



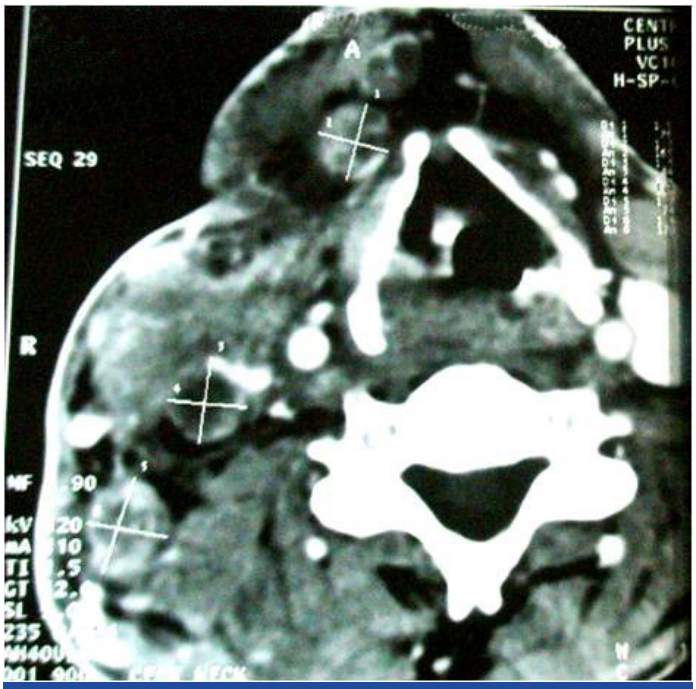
[Table/Fig-2]: Partial closure of right eye demonstrating facial nerve involvement



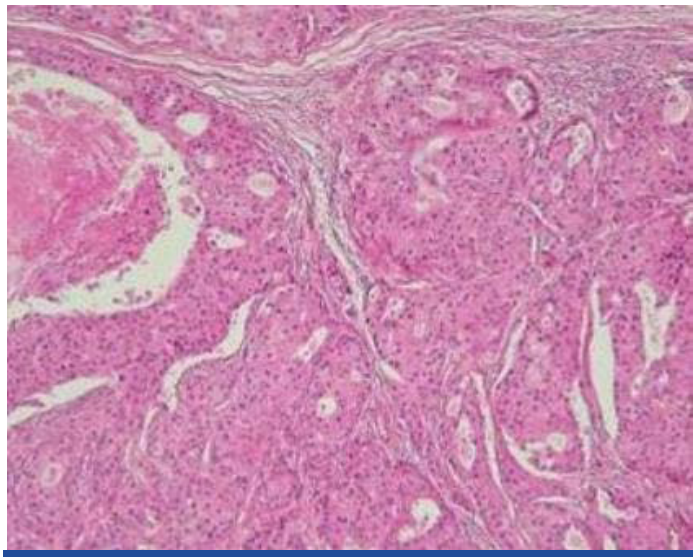
[Table/Fig-3]: Axial CT from the base of skull revealed diffusely enlarged parotid gland



[Table/Fig-4]: Axial CT revealed invasion of submandibular gland and sternocleidomastoid muscle



[Table/Fig-5]: Axial CT demonstrating involvement of ipsilateral lymph nodes

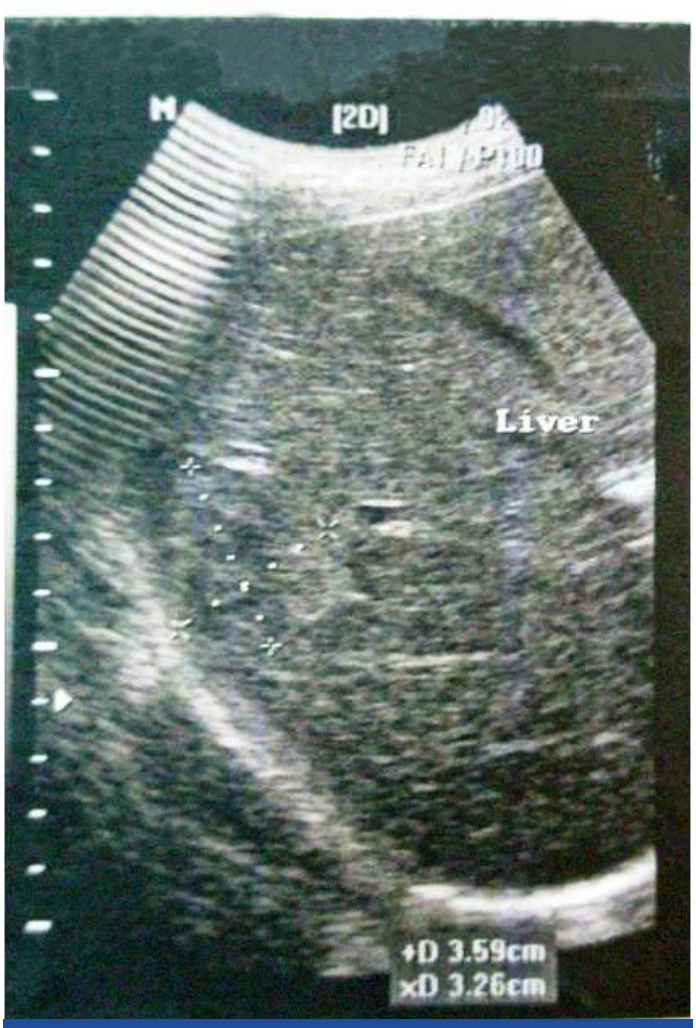


[Table/Fig-6]: H & E(40x) Areas of carcinoma component consisting epithelial clusters with necrosis in a chondromyxoid, fibrotic and sclerotic stroma

DISCUSSION

Although benign as many as 25% of pleomorphic adenomas can undergo malignant transformation if left untreated [1]. Carcinomatous transformation within a benign mixed tumour in which the initial benign elements are still identifiable is termed as Carcinoma Ex-Pleomorphic Adenoma (CXPA). The majority of CXPA develops from epithelial components of pleomorphic adenoma that display aggressive behavior [2]. The typical patient with CXPA has a long standing history of pleomorphic adenoma and suddenly experiences an increase in size, pain or facial nerve involvement. The CXPA is usually a more poorly circumscribed mass than the benign pleomorphic adenoma and is most likely to involve major salivary glands and typically occurs in 6th to 8th decade of life [3].

There are controversies on the treatment of CXPA in salivary gland. The incidence of positive margins, perineural invasion, facial nerve involvement and lymph node metastasis is higher than other malignancies of parotid gland. Post-operative adjuvant radiation could effectively eradicate residual deposits of microscopic disease. Currently, radical surgery with and without post-operative adjuvant radiation therapy is used [4,2]. CXPA is prone to recurrence and



[Table/Fig-7]: Ultrasonography abdomen revealed an ill defined focal lesion in postero-superior segment of right hepatic lobe

can metastasize. Prognostic markers are recurrence, capsular invasion > 9 mm and metastasis [5]. The development of associated metastatic disease is considered pre-terminal. In one series, disease specific survival was 45% at 3 years and 37% at five years, however, medial survival was 27% at 1 year after detection of any type of progression and 5% at three years after detection of distant metastasis [5].

It is reported that CXPA involves regional nodes with a frequency nearly equal to the distant metastatic rate. Olsen and Lewis reported that metastasis occurred regionally in 37 [56%] and distantly in 29 [44%] of 66 patients [5]. Malignant mixed tumours metastasize regionally and in distant areas, such as the lungs, hilar and cervical lymph nodes, bone and central nervous system [4-6].

Our case demonstrates a rare complication of parotid pleomorphic adenoma, malignant conversion followed by hepatic metastasis. Although local lymphatic spread is most common means by which these tumours metastasize, hematological distant metastasis have been reported to occur. Previously reported sites of hematogenous metastasis include the lung, pleura, kidney, ocular choroid and the brain and spinal cord [6,7]. In our case both regional lymphatic and distant hematogenous spread were identified. To our knowledge metastasis from a parotid CXPA to liver has not been reported in literature.

From a practical standpoint, however, the true importance of this case is that it poignantly illustrates the outcome that may result if there is a delay in diagnosis and treatment of benign pleomorphic adenoma. Because of the significant risk of malignant conversion, the presence of a pleomorphic adenoma should always be regarded as a premalignant condition and treated aggressively with appropriate surgery [2,4].

The fact that pleomorphic adenomas are classified as benign tumours should not overshadow the wide range of biological behavior associated with these neoplasms. When recurrence and distant metastasis occur, survival is remarkably low [5,6]. Therefore, early diagnosis and treatment of malignant mixed tumour of salivary gland is extremely crucial. Although metastasis to liver is rare, patients with carcinoma ex-pleomorphic adenoma should be investigated for metastasis to distant sites such as lungs, bones and liver.

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PARTICULARS OF CONTRIBUTORS:
1. Senior Lecturer, Department of Oral Medicine and Radiology, ITS Centre for Dental Studies and Research, Ghaziabad, Uttar Pradesh, India.
2. Senior Lecturer, Department of Pedodontics and Preventive Dentistry, SBB Dental College, Ghaziabad, Uttar Pradesh, India.
3. Senior Lecturer, Department of Oral and Maxillofacial Surgery, ITS Centre for Dental Studies and Research, Ghaziabad, Uttar Pradesh, India.
4. Senior Lecturer, Department of Oral Medicine and Radiology, ITS Dental College and Research Centre, Greater Noida, Uttar Pradesh, India.
5. Senior Lecturer, Department of Oral Medicine and Radiology, ITS Centre for Dental Studies and Research, Ghaziabad, Uttar Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Manu Dhillon,
Senior Lecturer, Department of Oral Medicine and Radiology, ITS Centre for Dental Sciences and Research,
Meerut-Delhi Road, Muradnagar, Ghaziabad, Uttar Pradesh, India.
Phone: 09219510522, E-mail: drmanudhillon@yahoo.co.in, drmanudhillon@gmail.com

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