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Metastasizing Pleomorphic Adenoma to Cervical Lymph Node: A Rare Case Report

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ABSTRACT

The metastasizing pleomorphic adenoma (MPA) is defined as histologically benign pleomorphic adenoma that exhibits local or distant metastases. It constitutes an extremely rare group of tumors. Most patients have a history of atleast one recurrence prior to the detection of local or distant metastasis. Although the exact pathogenesis remains elusive, hematogenous and lymphatic spread are proposed theories. Mortality in these cases can be as high as 22% thereby necessitating early diagnosis. Fine needle aspiration cytology serves as a minimally invasive and cost-effective tool in the diagnosis of this rare entity. We are presenting a case of recurrent pleomorphic adenoma metastasizing to lower cervical lymph node diagnosed on fine needle aspiration cytology.

Keywords- Metastasizing pleomorphic adenoma, cervical lymph node, fine needle aspiration cytology

INTRODUCTION

Metastasizing pleomorphic adenoma (MPA) is defined as histologically benign pleomorphic adenoma that exhibits local or distant metastases. [1] It is also known as metastasizing benign tumor. Though pleomorphic adenoma is the most common tumor of the salivary glands but metastasizing benign pleomorphic adenoma constitutes an extremely rare group of salivary tumors. [2] The most common initial tumor site is parotid gland (74%), followed by minor salivary glands (17%) and submandibular glands (10%). Most patients have a history of atleast one recurrence prior to the detection of local or distant metastasis. The most common site for metastasis is bone (45%), followed by head and neck (43%), lungs (36%), and abdominal viscera (10%). Within the head and neck area, only 17% of cases metastasize to regional lymph nodes. [3] The interval between the diagnosis of primary pleomorphic adenoma and its metastases varies from 3 to 52 years (mean 15 years) after occurrence of primary lesion. [1]

The exact pathogenesis is unknown but there are various proposed theories of which hematogenous spread is the most accepted one. It usually occurs after an incomplete surgical excision of the primary pleomorphic adenoma that favors seeding of the tumor cells in the lymphatics or blood vessels followed by the metastatic spread. It is the paradoxical nature of MPA, with its benign histological appearance and its metastatic spread capacity that has intrigued the clinicians and the pathologists worldwide. [2,4,5] We here present a case of metastasizing pleomorphic adenoma to cervical lymph node in a 38 year old female.

CASE REPORT

A 38 year old female presented with a painless left lower cervical swelling measuring 6x4.5x4 cm (fig 1). On local examination the swelling was non-tender, firm and situated deep to sternocleidomastoid.

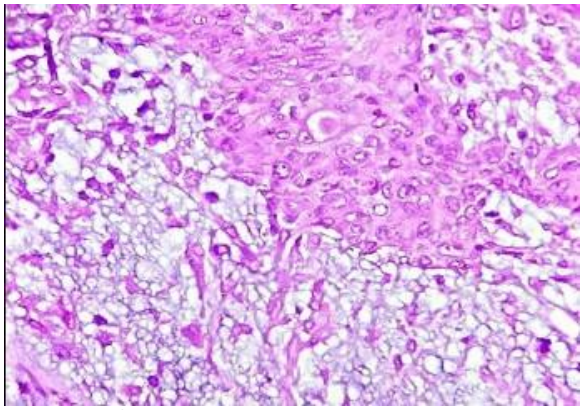

Figure 1- Left lower cervical swelling measuring 6x4.5x4cm

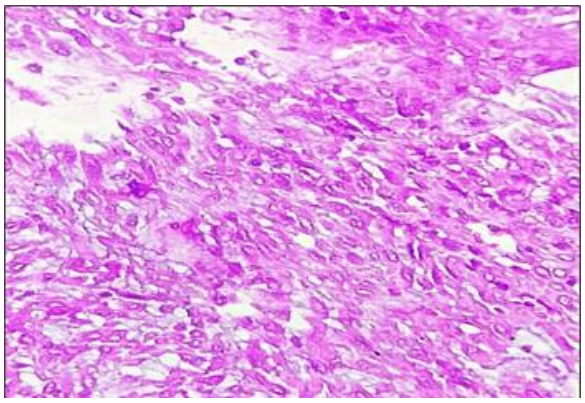

Figure 2- CECT showing large heterogenous lymph node mass in left lower cervical region

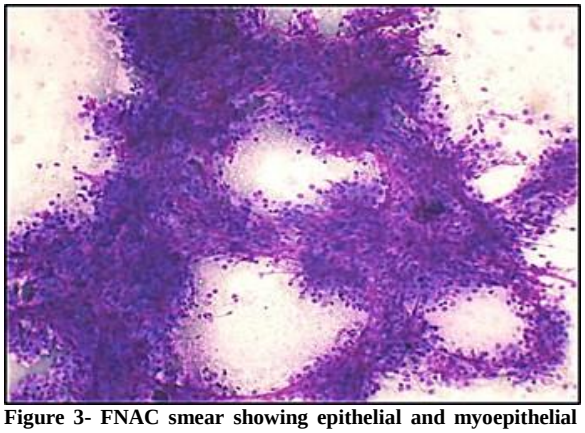

Figure 3- FNAC smear showing epithelial and myoepithelial cells embedded in abundant chondromyxoid stroma (Giemsa x 200)

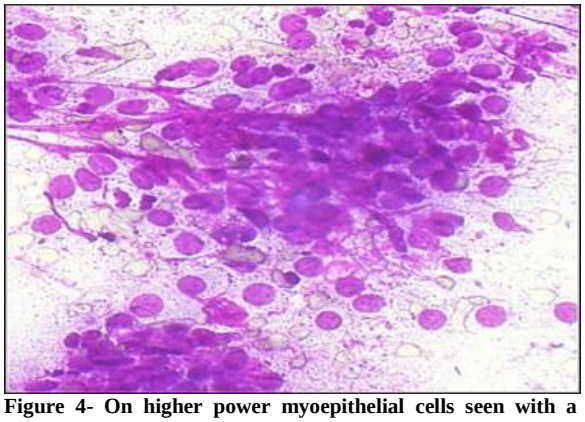

Figure 4- On higher power myoepithelial cells seen with a plasmacytoid appearance (Giemsa x 400)

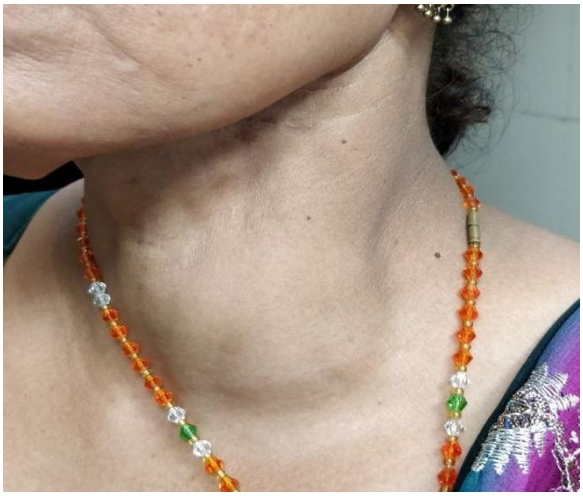

Figure 5- Section from previously excised submandibular gland showing biphasic population of benign epithelial islands and myxoid stroma (H&E x 200)

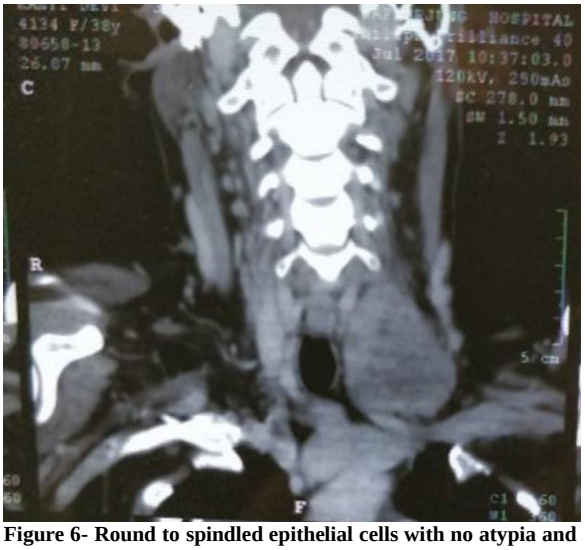

Figure 6- Round to spindled epithelial cells with no atypia and mitoses (H&E x 400)

CECT revealed the presence of a large heterogeneous lymph node mass in the left lower cervical region (fig 2). Patient had undergone two previous surgeries for recurrent benign pleomorphic adenoma in the left submandibular region two/three years back in the same hospital. Fine needle aspiration cytology (FNAC) from the lymph node swelling showed cellular smears comprising of dual population of epithelial and myoepithelial cells embedded in abundant chondromyxoid ground substance typical of a pleomorphic adenoma. There was no atypia, mitosis or necrosis (fig 3&4). No evidence of any lymphoid component was seen which suggested a complete effacement of the lymph node. Meanwhile

his previous histopathological slides from the submandibular swelling were retrieved from the department for a review. However, classical histological features of benign pleomorphic adenoma were seen (fig 5&6). Hence based on the past clinical history, histological and present radiological and cytological findings a final diagnosis of metastasizing benign pleomorphic adenoma was given. A complete excision for histopathological correlation was advised. However, the patient was lost to follow up.

DISCUSSION

Malignancy can arise from PA in three forms: carcinoma ex-pleomorphic adenoma (CEPA), carcinosarcoma and metastasizing pleomorphic adenoma (MPA). The latter two are exceptionally rare [6] with MPA accounting for 1% of all malignant PA. [7] It is histologically indistinguishable from pleomorphic adenoma but produces secondary tumors in distant sites. [1] To date 81 cases have been described. [8] The most common distant site is bone followed by head and neck and lung. Other sites include the kidneys, skin, liver, and brain. [8,9] Metastasis of a pleomorphic adenoma to the cavernous sinus is very rare, with only 1 case been previously reported. [10] The mechanism of MPA is poorly understood. It has been hypothesized to occur iatrogenically during surgical resection with seeding of disrupted tumor cells into the bloodstream. [11,12] Nourai et al. constructed a virtual series of 42 patients with MPA. Incomplete surgical excision of the primary PA was overwhelmingly associated with local recurrence and distant metastasis. This further supports the view that meticulous tumor resection with adequate margins should be conducted and enucleation avoided. [3] Czader et al. reported a case of MPA in which the patient presented with a solitary kidney tumor that showed the histological features of a PA in the absence of a previous or concurrent salivary gland neoplasm or salivary gland surgery. [4] Thirteen months after the removal of the kidney tumor, the patient presented an aggressive parotid tumor, which pathological examination yielded a CEPA. [4] These cases have challenged the classical theory of tumor seeding in blood vessels and lymphatics. He proposed that MPA and CEPA are different stages along a common biological pathway in malignant mixed-tumors spectrum and that metastasis capability of MPA more likely occurs secondary to accumulation of genetic mutations. [4] However, in our case the probable explanation is tumor cell seeding in the blood stream.

Knight et al showed that in patients who develop MPA the average age of PA presentation is 34.3 years with a range of 9 to 73 years, and the common decades of primary PA are the 2nd, 3rd and 4th. The most common decade was the 2nd. Therefore, younger presentation of PA could be a risk factor for developing MPA. [8]

Total surgical resection forms the mainstay of treatment. Adjuvant radiotherapy can be given to avoid missing hidden local recurrence that could eventually spread to distant sites. [2] Selective Neck Dissection (SND) and postoperative radiotherapy are indicated for regional lymph nodes metastasis. [13]

Prognosis is difficult to quantify for MPA as it is not widely reported. There is significant variability in disease free period and actual survival. However, poor prognostic factor includes multiple metastases, which are deemed invariably fatal. Often due to the moderate proliferation rate of MPA, it should be regarded and treated as an indolent malignancy, though other authors prefer the term low-grade malignancy, with prompt excision whenever possible. [14] Recurrence after complete surgical removal is rare, and the prognosis is excellent. [11]

CONCLUSION

Although rare, metastasizing benign pleomorphic adenoma must be kept in mind as a differential diagnosis in known cases of pleomorphic adenoma. It is a rare entity

which acts clinically as an indolent malignancy. There are no histological signs to predict MPA, though local recurrence after surgical excision is shown to be a risk factor. FNAC serves as a minimally invasive and cost-effective tool in the diagnosis of this rare entity.

Abbreviations

CECT- contrast enhancing CT scan
CEPA- carcinoma ex-pleomorphic adenoma
FNAC- fine needle aspiration cytology
MPA- metastasizing pleomorphic adenoma
PA- pleomorphic adenoma
SND- selective neck dissection

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