

Case Report

Clavicular Metastasis, an Initial Manifestation of a Malignant Pheochromocytoma - A Case Report

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Abstract

We present a 55 years old lady having an unusual presentation of abdominal pain and a clavicular mass as the initial manifestations of a nonfunctioning malignant pheochromocytoma. Fine needle aspiration of the clavicular mass revealed a metastatic adenocarcinoma. A search for primary site revealed a mass in left adrenal and multiple space occupying lesions in the liver. The aspirations were identical to that from the clavicle. Surgical biopsy of the clavicular mass revealed, features of metastatic pheochromocytoma. Urinary catecholamine was normal. Malignant pheochromocytoma can present with atypical features and guided fine needle aspiration cytology can be a safe diagnostic procedure.

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Key Words : Pheochromocytoma, malignant, non-functional.

Introduction

Pheochromocytomas are neoplasm derived from the chromaffin cells of adrenal medulla. The annual incidence varies between 1.6-2.1 cases per million. Although pheochromocytomas can occur at any age they are more common in fourth and fifth decades. More than 90% cases are sporadic with a female preponderance; the remainder affects younger males with a family history and as a component of multiple endocrine neoplasia (MEN). There is a classic 10 per cent rule for pheochromocytoma which indicates that 10% cases occur in children, 10% can be bilateral, 10% can be malignant and 10% can occur in extra-adrenal sites.

Case Report

A fifty five years old female patient presented with a swelling in right clavicular region. She had history of abdominal pain, vomiting, intermittent fever and loss of weight for the past two years. One year back she had cholecystectomy with a histopathological diagnosis of chronic cholecystitis. Her symptoms persisted even after the operation, followed by the appearance of a mass in the right shoulder of one month duration.

Conventional radiograph revealed an osteogenic lesion in the clavicle with calcification. Fine needle aspiration revealed a metastatic deposit of an adenocarcinoma. Abdominal ultrasound and computed tomography (CT) scan revealed a 7.5 x 8 x 10.6 cm solid mass in the left adrenal gland and multiple space occupying lesions (SOL) in the liver. Routine haematological tests showed a

raised erythrocyte sedimentation rate. Urinary catecholamines, VMA and metanephrines were all within normal range.

Image guided fine needle aspirations were done from the adrenal and liver SOLs. The smears were stained with May-Grünwald Giemsa (MGG) and haematoxylin and eosin stains. The aspirates from the adrenal and liver SOL were cytologically similar to that from the clavicular mass. The smears were highly cellular with cells arranged in loose clusters and also dispersed in the background. The cells were polygonal in shape having abundant, fragile cytoplasm and red granularity in MGG stained smears. The nuclei were eccentrically placed. An unusual feature was vacuolation in the cytoplasm giving a signet ring type of appearance (Fig. 1).

The differential diagnosis was metastasis from an adenocarcinoma arising in lung, breast, intestine or kidney. Ultrasound, computed tomography scan and endoscopy could not detect any primary tumour in these sites. Another possibility was adrenocortical carcinoma with metastasis, which was discarded once the clavicular mass was histologically diagnosed as metastatic pheochromocytoma (Fig. 2).

The cytological diagnosis was malignant pheochromocytoma with hepatic and clavicular metastasis. This was followed by a surgical biopsy of the clavicular mass which confirmed the diagnosis by histology. As the adrenal mass was surgically unresectable only palliative treatment could be given to the patient.

Discussion

Pheochromocytoma is the paraganglioma of adrenal medulla arising from the chromaffin cells. The classical clinical symptoms are tachycardia, headache, sweating and paroxysmal hypertension. These symptoms are produced by the intermittent excessive

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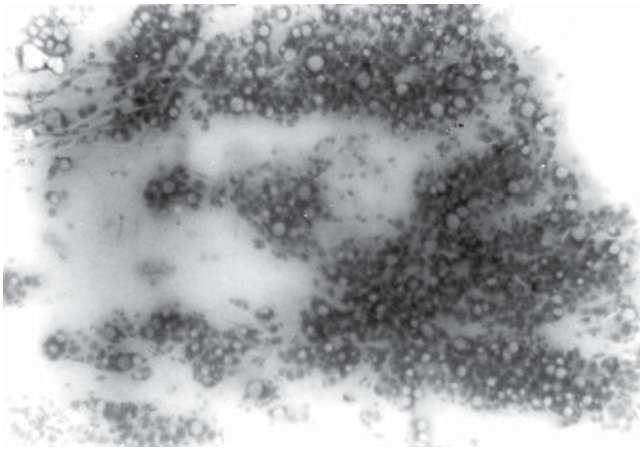


Fig. 1 : Smear from clavicular mass showing loose sheets of cells with eccentrically placed nucleus and cytoplasmic vacuoles (H&E, x 265).

secretion of catecholamines from the chromaffin cells. This is also accompanied by the elevation in the level of the serum and urinary catecholamines and their metabolites.

Our patient never had any cardiovascular symptoms. Her blood pressure was also within normal range for her age. She had only abdominal pain, vomiting, intermittent fever and weight loss. The catecholamines were also within normal range. In a recent study it has been indicated that the classical symptoms of pheochromocytoma are more common in benign tumours whereas abdominal pain and dorsalgia being more frequent in the malignant variety.¹ Although rare, as many as 20% of pheochromocytomas can be nonfunctional.^{2,3} In these cases the patients remain asymptomatic, the biochemical findings are non-diagnostic and medical intervention is invariably delayed till the appearance of a metastasis.

The cytological and histological features are often identical in benign and malignant pheochromocytomas. It is only the presence of invasion or metastasis which confirms the malignant nature of the lesion.^{4,5} Therefore all the cases of pheochromocytomas should be carefully searched for any metastatic deposit. As the malignant pheochromocytomas have a predilection for skeletal metastasis often with the exclusion of other organs, patients with a suspected pheochromocytoma should have a preoperative bone scan.⁶

The primary treatment for pheochromocytoma is surgical resection. The prognosis becomes unfavourable in late tumour detection and for malignant tumours. So to avoid delay in treatment it is important to remember that pheochromocytomas

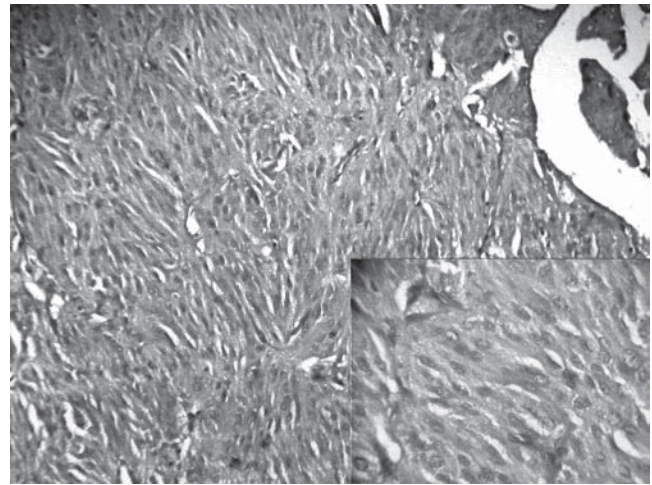


Fig. 2 : Section from clavicular mass showing tumour cells in Zellballen pattern (H&E, 300). Inset shows nuclei with prominent nucleoli.

can be asymptomatic or can present with atypical features, and biochemical tests may be non-diagnostic. In nonfunctional tumours with inconclusive biochemical tests, image guided fine needle aspiration of adrenal mass can be used as a safe diagnostic procedure.

As the histological criteria are not enough to confirm the benign nature of a pheochromocytoma, all patients should be carefully followed up after surgical resection to detect any recurrence or metastasis.^{7,8} In other words all cases of pheochromocytomas should be treated as potentially malignant.

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