IJNRD.ORG ISSN : 2456-4184
INTERNATIONAL JOURNAL OF NOVEL RESEARCH AND DEVELOPMENT (IJNRD) | IJNRD.ORG
An International Open Access, Peer-reviewed, Refereed Journal

PIGMENTED PHEOCHROMOCYTOMA OF ADRENAL MEDULLA

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INTRODUCTION

Pigmented pheochromocytoma is an extremely rare variant of pheochromocytoma, a neuroendocrine tumour arising from adrenal medulla. Only 16 cases have been reported in literature till date (1-7). The nature and origin of the pigment seen in these tumours is uncertain, being variably described as Lipofuschin, melanin or neuromelanin. We present a case of this unusual tumour in a 45year old female patient, who presented with pain abdomen since 1year.

CASE REPORT

A 45year old women presented with complaints of abdominal pain since 1year. Her blood pressure and systemic examination were within normal limits. Abdominal ultrasound revealed a left suprarenal multiloculated, cystic mass measuring 9*4.5cm. Computed tomography showed a large, ill-defined mass measuring 9.8*4.7cm in left suprarenal area with solid and cystic components. Urinary vanillylmandelic acid was within normal limits. There was no family history of multiple endocrine neoplasia. The patient underwent left adrenalectomy with resection of the mass. The post operative course was uneventful.

PATHOLOGICAL FINDINGS

Grossly, the specimen was well encapsulated yellowish and blackish mass measured 10*5*3cm and was partly cystic and partly solid on cut surface (fig. 1&2).

Microscopically showed tumour composed of round to polygonal cells arranged in organoid and Zellballen pattern with thin-walled sinusoidal blood vessels separating the cell clusters. The cells

showed vesicular to pleomorphic nucleus with a moderate amount of granular, eosinophilc cytoplasm(fig.3&4). At places, abundant, coarse, brown-black melanin pigment is seen in the cells(fig.5&6).There was no evidence of necrosis or mitotic activity. Thus the morphologic diagnosis is pigmented pheochromocytoma was made. On immunohistochemistry, tumour cells were immunopositive for chromogranin(fig.7) and synaptophysin(fig.8), while they are negative for HMB-45(FIG.9) and S-100(fig.10), ruling out the possibility of malignant melanoma. Based on the histomorphological features and immunohistochemistry, a final diagnosis of pigmented pheochromocytoma was made.

DISCUSSION

Pigmented pheochromocytoma are extremely unusual adrenal tumours. 16 cases have been reported in literature till date, as single reports or as a part of series on pheochromocytoma .

Most reported patients with this neoplasms were females and some of reported cases were associated with syndromes like multiple endocrine neoplasm(MEN), neurofibromatosis(NF), and Von Hippel Linau disease(VHL). Other cases were noy associated with any of these syndromes.

Identification of broen to black pigment in an adrenal tumour can lead to a diagnostic dilemma. The differential diagnosis of pigmented pheochromocytoma includes other pigmented neoplasms that may be seen in the adrenal gland such as malignant melanoma, either primary or metastatic and pigmented adrenal adenoma also known as “black adenoma”

Metastatic MM is a likely possibility in a patient with bilateral adrenal masses. Malignant melanoma arising primarily in the adrenal or a metastasis is associated with a rapidly progressive clinical course(8)

In our case tumour cells lacked prominent eosinophilic nucleoli characterstic of MM, were immunopositive for chromogranin and synaptophysin and were immunonegative for S-100 and HMB-45, thus ruling out MM. Adrenal cortical adenomas with a brown to black appearance due to black adrenal adenomas.(BAs)

Microscopically, BAs are composed of polygonal cells arranged in sheets or cords, quite similar to pheochromocytoma. However a portion of tumour cells contain pale, lipid-rich cytoplasm and some of the tumour cells granular brown to golden-brown pigment.

The histogenesis of pigment within a phelochromocytoma remains obscure. most authors propose the theory that are both the adrenal medulla and melanocytes orginate from the neural crest, the presence of melanin in the adrenal medullar neoplasm can be explained by divergent differentiation of neural crest cells.

Another hypothesis proposed by Belleza et al, is that degradation of cetacholamines in a pheochromocytoma leads to production of neuromelanin, a lipofuschin-like pigment. Our case is more in line with the former hypothesis.

CONCLUSION

Pigmented pheochromocytoma is a very rare tumour. We report the seventeenth case of pigmented pheochromocytoma.

Although the presence of pigment in the tumour does not affect the clinical course, it is important from the differential diagnostic point of view, and other pigmented tumours that occur in the adrenal, like malignant melanoma, either primary or metastatic and black adenomas need to be excluded.

Histochemistry, immunohistochemistry and electron microscopy are of great value in distinguishing between these lesions.

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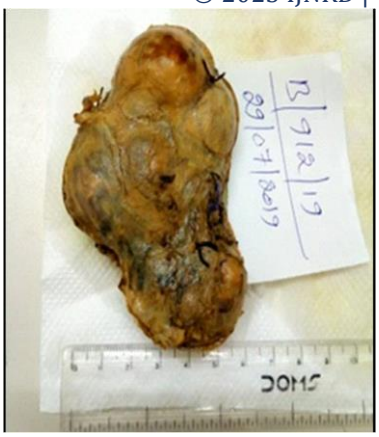

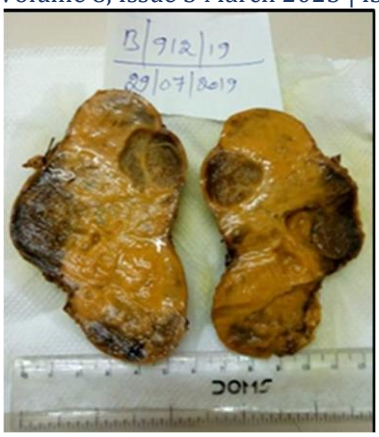

Fig 1- Gross of tumor

Fig 2- Cut section of tumor

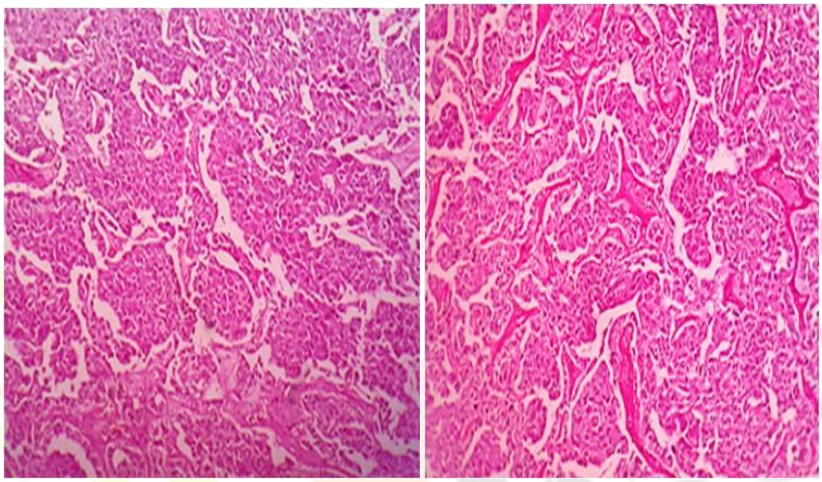

Figure 3 & 4:Tumor cells showing Zellballen pattern 400X

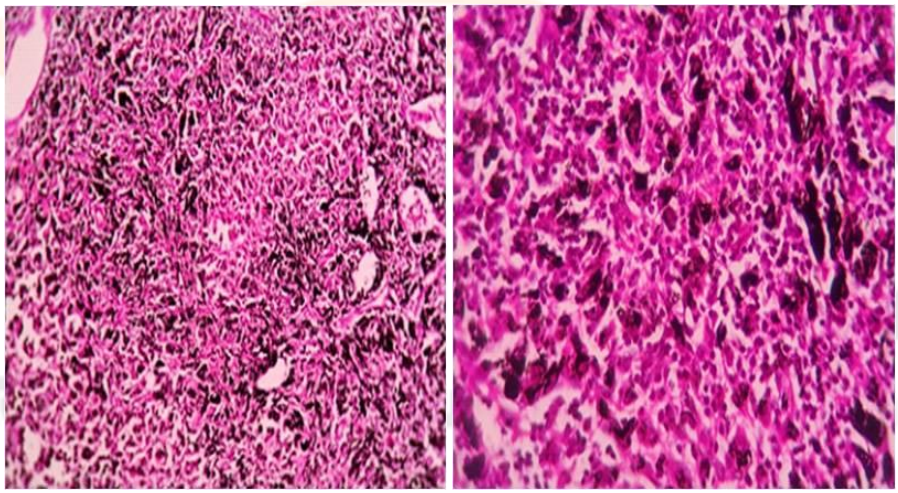

Figure 5 & 6:Tumor cells showing brown black pigment 400X and 1000x

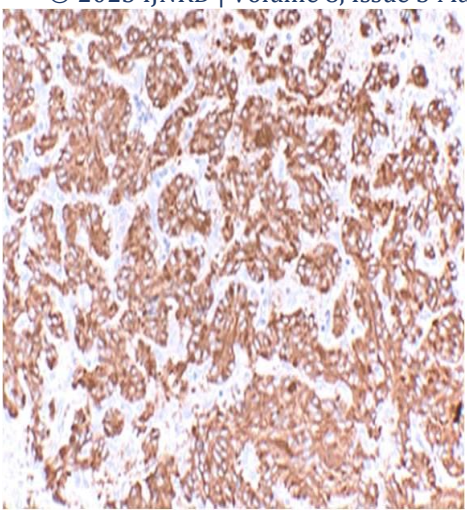

Fig 7- Immuno-positivity for chromogranin 400X

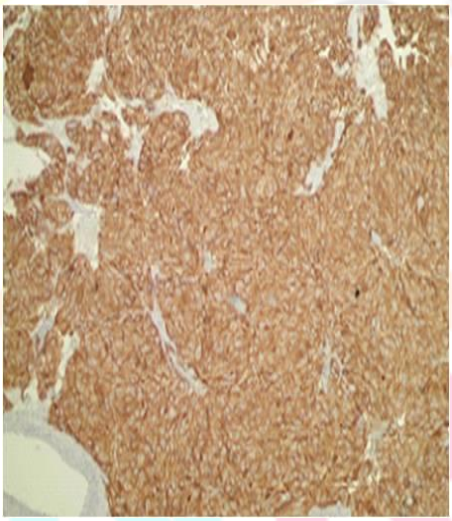

Fig 8- Immuno-positivity for Synaptophysin 400X

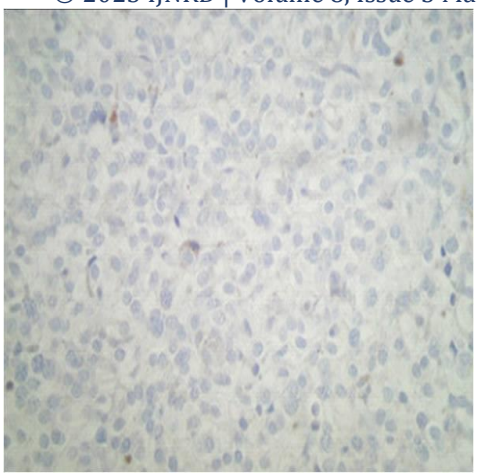

Fig 9- Immuno-negativity for HMB 45 1000X

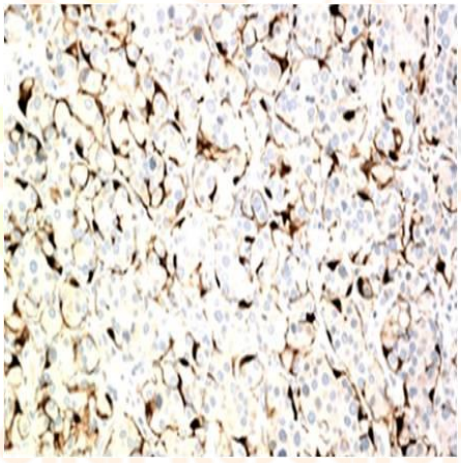

Fig 10- Immuno-neagtivity for S-100 1000x

Legends to figures:

Figure 1:Gross of tumor

Figure 2:Cut section of tumor

Figure 3 & 4:Tumor cells showing Zellballen pattern 400X

Figure 5 & 6:Tumor cells showing brown black pigment 400X and 1000x

Figure7:Immuno-positivity for chromogranin 400X

Figure 8:Immuno-positivity for Synaptophysin 400X

Figure 9:Immuno-negativity for HMB 45 1000X

Figure 10:Immuno-neagtivity for S-100 1000x