

## Primary Malignant Melanoma of Adrenal Gland

Usman Hassan<sup>1</sup>, Mudassar Hussain<sup>2</sup>, Hamna Salahudin<sup>3</sup>, Sheeba Ishtiaq<sup>4</sup>, Noreen Akhtar<sup>5</sup>

<sup>1,2,5</sup> Consultant Histopathologist, Shaukat Khanum Memorial Cancer Hospital and Research Centre, Lahore

<sup>3</sup> Resident Histopathology, Shaukat Khanum Memorial Cancer Hospital and Research Centre, Lahore

<sup>4</sup> Assistant professor of histopathology and Consultant Histopathologist Gulab Devi Hospital, Lahore

### ABSTRACT

A 49 years old male presented with abdominal pain for two months. Abdominal examination revealed slight tenderness in the right lumbar region. On radiological examination an abnormal lesion, measuring 5.5cm x 4.5cm, involving the right adrenal gland was found. It was biopsied and histological and immunohistochemical features were consistent with malignant melanoma. After meticulous physical examination, the patient did not have ocular or muco-cutaneous melanoma and he did not have any past significant medical or surgical history. We labelled this case as primary malignant melanoma of adrenal gland. Unfortunately, the patient died two months after the detection of tumor and before the start of treatment.

**Key Words:** Abdominal pain, Adrenal mass, Malignant melanoma

#### Address of Correspondence

Usman Hassan

Email: drusmanhassan256@gmail.com

#### Article info.

Received: March 24, 2018

Accepted: May 12, 2018

**Cite this case Report:** Hassan U, Hussain M, Salahudin H, Ishtiaq S, Akhtar N. Primary Malignant Melanoma of Adrenal Gland. 2018; 7(2):154-156.

**Funding Source:** Nil

**Conflict of Interest:** Nil

### Introduction

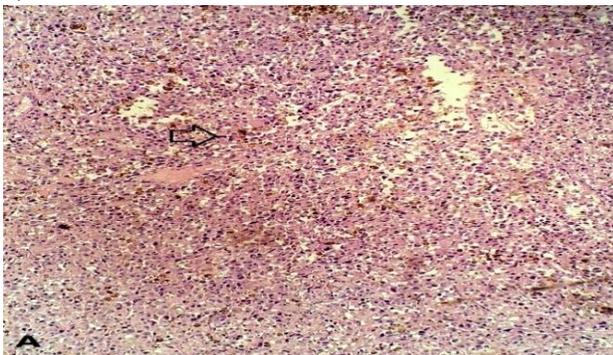
Malignant melanoma is a neoplasm of skin. Apart from the skin and choroid plexus, primary malignant melanoma at other sites is a rare entity.<sup>1</sup> Documented sites where malignant melanoma can occur are lung<sup>2</sup>, genitourinary tract<sup>3</sup>, and GIT<sup>4</sup>. It is very rare in adrenal gland. Only 12 well-documented cases of primary malignant melanoma of adrenal gland have been reported.<sup>5</sup> In the adrenal gland it originates from neural crest cells present in adrenal medulla.<sup>6</sup> It is difficult to differentiate between primary and metastatic malignant melanoma in adrenal gland. Histological and immunohistochemical studies are not helpful in differentiating between primary and metastatic malignant melanomas.<sup>6</sup> Therefore meticulous physical examinations is necessary to rule out the possibilities of primary ocular or mucocutaneous melanomas. Up till now, primary malignant melanoma of adrenal gland has not been reported from Pakistan.

### Case Report

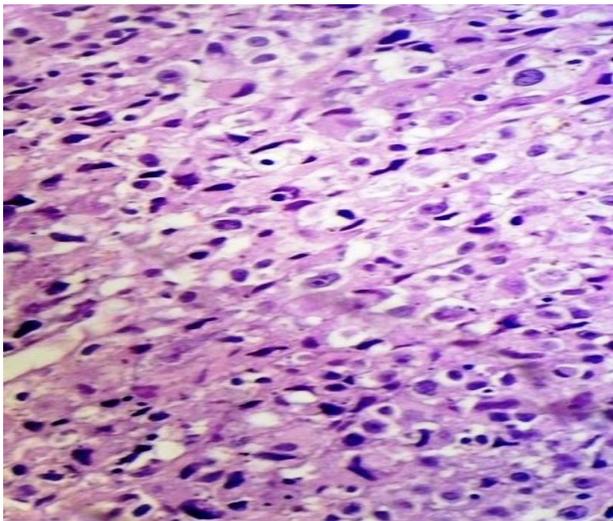
A 49 years old male presented with the complaints of abdominal pain, anorexia and weight loss for two months. He did not have any significant past medical or surgical history. On general physical examination patient was emaciated and pale. Abdominal examination only revealed slight tenderness in right lumbar region. Laboratory investigations showed haemoglobin of 10.0 g/dl with MCV and MCHC 78.6 fl and 30.6g/dl respectively. Renal function tests, liver function tests, and electrolytes were normal. Abdominal ultrasound showed a solid hypoechoic mass of 5.5cm x 4.5cm, above the upper pole of right kidney. It did not show increased flow on Doppler. Left kidney, liver, gall bladder, bile duct, spleen and pancreas were normal. No pleural effusion, ascites or para aortic lymphadenopathy was noted. 3D CT abdomen with IV contrast revealed a dense abnormal lesion, measuring 5.5cmx4.5cm, involving the right adrenal gland. Both kidneys showed multiple cysts with no solid mass or obstructive changes. Liver, gall bladder, spleen,

pancreas, aorta and IVC were normal. No ascites was seen.

Patient underwent diagnostic laparoscopy and an incisional biopsy was taken from right adrenal mass. On gross examination, we found multiple fragments measuring 2.0cm x 1.0cm x 1.0cm in aggregate. Specimen weighed 8 grams. Cut surface revealed hemorrhagic and necrotic areas. Microscopic examination revealed a malignant neoplasm, comprising of polygonal cells arranged in diffuse sheets. The cells showed marked pleomorphism, atypia, atypical mitoses (11/10 HPF) and few cells showed intracytoplasmic melanin pigment (Fig 1). Few areas showed necrosis.

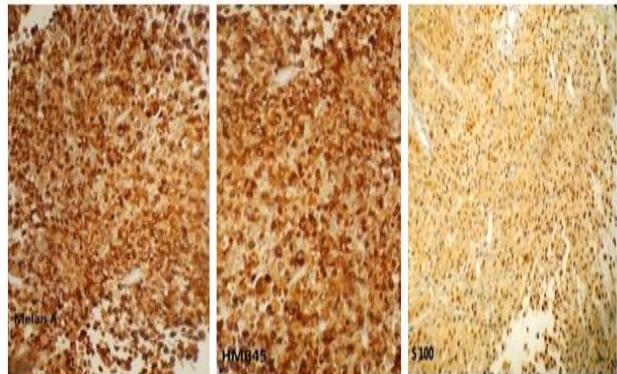


**Figure 1(A):** Low power view shows sheets and clusters of medium to large sized atypical polygonal cells with focal areas of pigmentation (arrow) [H&E 10X100].



**Figure 1(B):** High power view shows atypical tumor cells with rounded vesicular nuclei, prominent nucleoli, intra cytoplasmic melanin pigment [H&E 40X100].

Tumour cells revealed positive expression for Melan A, S100, HMB-45 (Fig 3\2) and negative staining for synaptophysin, calretinin, inhibin, Cytokeratin, LCA.



**Figure 2: Positive immunohistochemical staining for MELAN A ,HMB-45 AND S100.**

Based on above-mentioned results, an initial diagnosis of malignant melanoma was made and patient was evaluated for any possible primary site of malignant melanoma. On meticulous evaluation, physicians did not find any ocular or mucocutaneous melanoma. Patient was diagnosed as case of primary malignant melanoma of adrenal gland. He was referred to oncologist for further management. However, patient died before the start of treatment, two months after diagnosis of adrenal malignant melanoma. Autopsy was not performed due to lack of attendant's consent.

## Discussion

Malignant melanoma comprises of 2% of all body tumours.<sup>7</sup> Primary and metastatic melanomas of the adrenal gland are rare and are incidental findings. Pain is the most common manifestation along with gastrointestinal symptoms caused by the compression of the structures adjacent to the tumour.<sup>8</sup> Primary melanoma of the adrenal gland is usually a voluminous, non-functional tumor showing heterogeneous contrast enhancement on the computed tomographic (CT) scan and diagnosis is made on the basis of immunohistochemical studies and criteria led by Carstens et al.<sup>2</sup> Adrenal glands can be the sites of metastatic deposits from cutaneous or visceral melanomas in up to 50% of cases.<sup>9,10</sup> The presence of melanoma in both adrenal glands favors metastatic melanoma over a primary adrenal melanoma.<sup>11</sup>

Histopathological and immunohistochemical studies cannot usually differentiate between primary and metastatic malignant melanomas. Primary melanoma of adrenal gland is a very rare entity.<sup>12</sup> Only 12 cases have been reported to date.<sup>13</sup> It is important to rule out possibilities of adrenocortical carcinoma, pheochromocytoma and metastatic melanoma before a diagnosis of primary malignant melanoma of adrenal gland is made.

The patients presented by Parker and Vincent et al,<sup>12</sup> Kinseley and Baggentons et al,<sup>14</sup> Dick et al<sup>15</sup> Sasidharam et al,<sup>16</sup> Granero et al<sup>17</sup> and Luis González-Sáez et al had more than 50 years of age while our patient was 49 years old similar to the cases presented by Dao et al, Liatsikos et al, and Zalatnai et al respectively.<sup>6,18,19</sup> The previously reported cases as well as our case had a tumor size of more than 5 cm. The largest reported adrenal tumor measured 17cm x 12cm x 10cm. The cases presented by Kniseley and Baggentoss et al,<sup>14</sup> Parker and Vincent et al,<sup>12</sup> Dao et al<sup>6</sup> and Zalatnai et al,<sup>19</sup> died shortly after detection of disease. Our patient also died after two months of detection. This indicates poor prognosis of the disease.

In one study of 31 patients who underwent adrenal surgery for metastasis at a single institution over 10 years' period (1999-2008), the primary tumor diagnosis was non-small cell lung carcinoma in 20, colorectal carcinoma in 5, renal cell carcinoma in 2 and malignant melanoma and breast carcinoma in one case each.<sup>20</sup> In our case there was a unilateral adrenal involvement. There was no ocular or muco-cutaneous melanoma. This highly favors primary malignant melanoma of adrenal gland. Out of 279929 samples processed at our institute for various diseases during last five years, 429 cases were diagnosed as malignant melanoma. Only one out of these 429 cases was adrenal malignant melanoma.

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