RIGHT RENAL MASS, A RARE DISEASE AND DIAGNOSTIC DILEMMA IN A YOUNG FEMALE

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Abstract

A 22-year-old female from Afghanistan presented with history of pain right lumbar region along with feeling of heaviness and weight loss. The abdominal examination revealed a mass approximately 10*10 cm in the right lumbar region which was bimanually palpable. The diagnosis of Right lumbar mass was made on ultrasound. Final CT scan confirmed its origin from right kidney along with metastasis in lung. The ultrasound guided biopsy turned out to be spindle cell tumour of kidney. Right sided nephrectomy along with removal of proximal ureter done. Histopathology confirmed the diagnosis of mesenchymal chondrosarcoma of right kidney. Post-operative outcome of patient was uneventful.

Key Words: Extra skeletal Mesenchymal Chondrosarcoma, Renal Extra skeletal Mesenchymal Chondrosarcoma.

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Mesenchymal chondrosarcoma is a very rare variety of chondrosarcomas. It represents 2% of the chondrosarcomas and is one of the rarest forms of tumour of extra skeletal origin.[1] Only few cases arise from the kidney.[2] There are only nine cases reported previously according to the literature.[3] According to the available data, our case is 10th one. This case was a diagnostic dilemma. The patient remains undiagnosed till she was operated.

Case Presentation

A young female 22 years of age presented with history of pain and fullness in right lumbar region. The fullness kept on increasing in size for 2 years. There was a feeling of heaviness and weight loss. She was complaining of loss of appetite and backache. There is no history of nausea, vomiting, diarrhea, hematuria, and cough with expectoration. The rest of systemic history was unremarkable. On examination, patient was thin lean with mass of 12*10cm palpable in right lumbar region. The mass was bimanually palpable and firm to hard in consistency. The mass was non-tender and not mobile. There were no signs of intestinal obstruction. Rest of the systemic examination was unremarkable. On the basis of this history and examination, the differential diagnosis was Renal cell carcinoma or lymphoma. Her hemoglobin was 8g/dl, rest of her hematological workup was unremarkable. Her Renal function tests were also within normal limits. On ultrasound abdomen, the mass originated from right kidney and was 10×8 cm in size. CT Scan abdomen done which showed, Large well defined multiseptated lesion 12×11×8 cm was noted in Right lumber region with multiple regional lymph nodes (fig, 1) There were soft tissue nodules in Right and Left lungs along with pleural effusion (fig 2). There were multiple sclerotic lesions in iliac region which highly suggested of metastatic disease.

Ultrasound guided trucut biopsy done and the report of spindle cell carcinoma came. The report was rechecked from another laboratory which came out

Figure. 1: CT Scan abdomen showing mass.

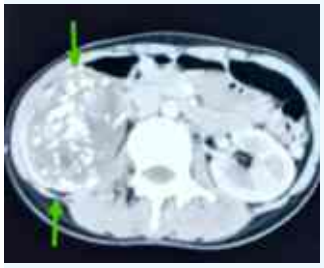
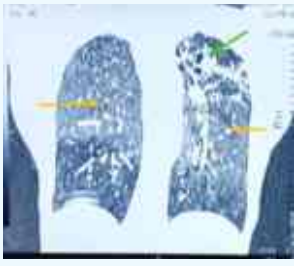


Figure. 2: CT chest showing nodules in lung



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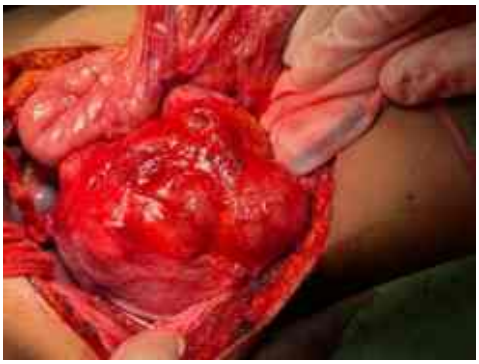
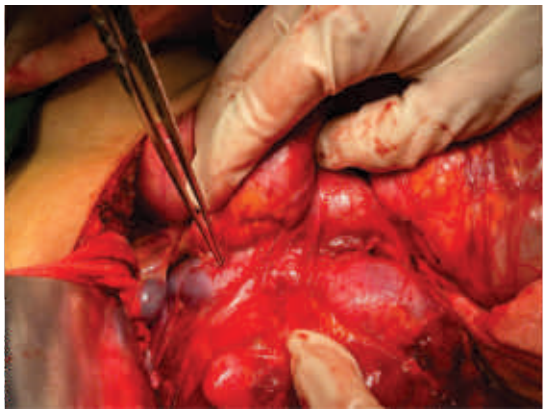
to be Ewing's sarcoma (round blue cell tumour). The patient received 12 cycles of chemotherapy from Afghanistan which she had no record available. The CT scan was done again which showed no change in size of tumour. Final diagnosis of spindle cell carcinoma was made and exploration planned finally. Renal scan was done to confirm the normal functioning of left kidney. Patient was evaluated by anesthetist; her hemoglobin was built along with her pulmonary and cardiac fitness taken. Bed was arranged in intensive care unit for her. Right sided nephrectomy along with proximal ureterectomy was done. The operative Findings were 16×10×7cm large renal mass involving the adrenal gland (Fig 3). Renal vein and artery. Inferior Vena Cava was spared(Fig 4). No para aortic para caval lymph node. Rest of abdominal visceras were normal.

Under aseptic measures right subcostal incision given. Skin and subcutaneous tissue excised. External oblique, internal oblique and transverses abdominus excised. Peritoneal cavity opened. Above mentioned findings noted duodenal kocherization done and retroperitoneal space aproached. Hilum of kidney identified. Renal artery and vein ligated (Fig 5). Tumor separated from I VC adrenal vessels ligated. Proximal ureter ligated. Tumor separated from surrounding visceras and resected. Hemostasis secured. Closure of abdomen in layers done and a subcostal drain was placed. Final specimen retrieved (Fig 6).



Right renal mass

Figure 3: *Inferior Vena Cava Along with Mass*



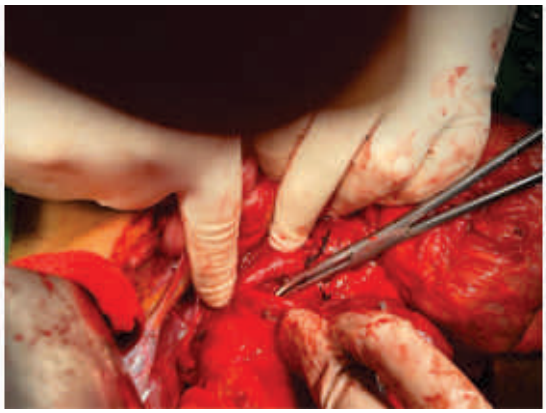


Figure 4: *Right Renal Vessels*

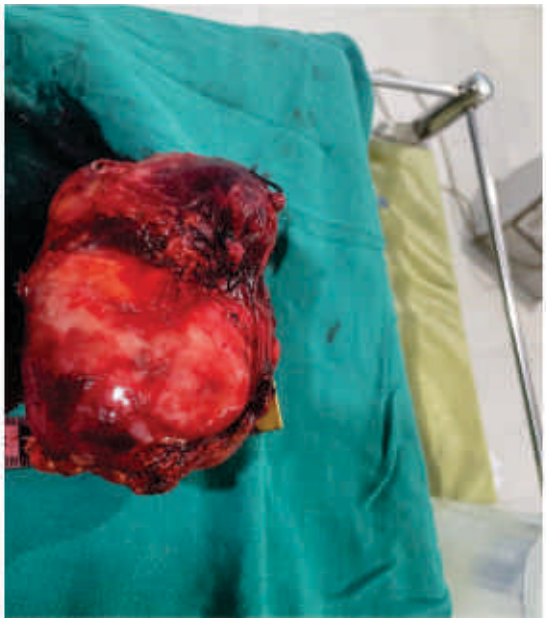


Figure 5: *Final Specimen*

Figure 6:

Final histopathology report came out and final diagnosis of Mesenchymal Chondrosarcoma of right

kidney with all of its margins clear was made. Patient was discharged in a stable condition.

DISCUSSION

Mesenchymal Chondrosarcoma is a rare disease making 2% of the chondrosarcomas.[4] Extra skeletal Mesenchymal chondrosarcoma, due to the rarity of this disease and variable presentation, its diagnosis is also a challenge and may lead to a misdiagnosis initially.[5] Mesenchymal Chondrosarcoma is more common in males then females and its other way round with extra skeletal mesenchymal chondrosarcoma.[6] Huvos et al recommended in his study about the guidelines for the treatment of skeletal chondrosarcomas.[7] Knott et al described the recomendations for extra skeletal mesenchymal tumours treatment. The better option is surgical complete excision with all margins clear.[8] The role of chemotherapy and radiotherapy is not established in EMC of kidney as it is very rare tumour. There is established role of Chemotherapy and Radiation in EMC which improves the overall survival. Bishop et al explained the comparison of treatment of EMC with chemotherapy and radiation. According to them there is overall improvement of survival with adjuvant chemo/Rad, as compared to surgery alone. They never mentioned about the EMC of kidney.[9] Gherman V et al, discussed the management of a case of extraskeletal mesenchymal chondrosarcoma of a kidney in a female. According to them it is metastatic disease and needs excision with complete margins as compared to our case report. This is also very rare tumour and only 9 are reported previously with a very poor outcome and managed without chemotherapy and radiotherapy.[10] Another case report by Valente P et al, narrated that EMC of renal origin is very rare disease with bad prognosis and poor outcomes.[8] This tumour is so rare that in another case report patient passed away after 8 months of resection.[11] It is highly metastatic tumour which has a very poor outcome even with chemotherapy.[12] According to the latest literature update 16 cases of Renal EMC are documented and after including our case it's 17.[11]

CONCLUSION

Renal extraskeletal mesenchymal chondrosarcoma is one of the rare disease. The surgical resection of the tumour with clear margin is the most optimal treatment. It is metastatic disease with poor prognosis. The role of chemotherapy and radiation is yet to be established.

Conflict of Interest *None*

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