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A RARE TUMOR – ADRENAL ANGIOSARCOMA (case report)

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Background. *Primary adrenal angiosarcoma is an exceptionally rare tumor with an incidence of <1% of soft tissue sarcomas. Less than 50 cases being described in literature; most of them were nonfunctional presenting with symptoms of abdominal lump, weight loss, fatigue or weakness. The treatment remains multi-modal with involvement of surgeon as well as medical oncologist.*

Objective. *The aim of this case report is to describe an entity which is rare in terms of incidence. It was functional as well as metastatic and managed with multimodality therapy.*

Methods. *The case is a 47-year-old man diagnosed with metastatic adrenal angiosarcoma. The patient was managed with chemotherapy followed by surgery.*

Results. *The final histopathological examination of the specimen and immunohistochemistry revealed angiosarcoma. At present the patient is under follow-up.*

Conclusion. *Even though surgery forms the main stay of management, the role of multi-modality therapy forms the cornerstone for adrenal angiosarcomas.*

KEYWORDS: **adrenal angiosarcoma; suprarenal; adrenalectomy; vasoformative tumor.**

Introduction

A tumor with an incidence of <1% of soft tissue sarcomas, angiosarcoma is a rare lesion arising out of vascular endothelium [1, 2]. Primary angiosarcomas of visceral organs and bones are exceedingly rare, though they account for metastatic sites. Most of the primary lesions are either from the skin or subcutaneous tissue [3,4]. Out of this meagre incidence of primary visceral angiosarcomas, adrenal angiosarcomas constitute an exceptionally uncommon variety with less than 50 cases reported in literature [5]. Hence this case report becomes important and unique due to the rarity of the tumor. This case depicts the management of a 47-year-old man with meta_ static left adrenal angiosarcoma.

Case report

A 47-year-old man with no previous comorbidities initially presented with blurring of vision of one-month duration. His initial clinical examination revealed a blood pressure of 225/150 mm of Hg for which he was diagnosed as hypertensive emergency with retinopathy and treated with antihypertensives. Assessment of his hematological and biochemical parameters revealed an abnormal serum urea 26.9 mg/dL, creatinine 2.14 mg/dL and uric acid 9.5 mg/dL. He was further evaluated for cause of the same and was found to have a left suprarenal mass measuring 15×18×22 cm extending on to the peripancreatic region on abdominal sonography. Further imaging with MRI of the abdomen revealed a large suprarenal mass lesion measuring 13×17×18 cm with fat, soft tissue areas and calcification with inferior displacement of kidney and loss of fat planes with left dome of diaphragm, anterior and posterior renal fascia. In addition, multiple soft tissue deposits were found along anterior abdominal wall, largest measuring 4.5 cm along left hypochondrium. Multiple retroperitoneal lymph nodes were also enlarged.

Whole body PET CT revealed diffuse FDG uptake in left suprarenal lesion of similar size along with multiple nodules in both lungs with FDG uptake in paratracheal lymph nodes. FDG uptake was also noted along the anterior abdominal wall in the left hypochondrium. Ultrasound guided fine needle aspiration cytology from left adrenal mass lesion as well as anterior abdominal wall deposits revealed a malignant vasoformative tumor with cells having large hyperchromatic nuclei and abundant vesicular cytoplasm. These cells stained positive for ERG, CD34 and CD31 on IHC. Patient was diagnosed as a case of metastatic adrenal angiosarcoma based on the imaging as well as tissue biopsy findings.

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Patient was started on chemotherapy based on Paclitaxel and administered six cycles. Follow up PET CT after 6 months of initial presentation revealed a relatively stable disease with no newer lesions. However, there was no regression in size of the lesions. In view of the same, patient was referred for left adrenalectomy. He underwent left open adrenalectomy along with splenectomy. Intra-operatively 15×15 cm highly vascular tumor was found with dense adhesions between spleen, greater curvature of stomach and tail and body of pancreas with involvement of splenic artery by the tumor necessitating a splenectomy. Left kidney was free of tumor. Patient had an eventful post-operative course with surgical site infection requiring frequent dressings for a period of two weeks and secondary closure. He was administered requisite vaccines against OPSI in the post-operative period.

Histopathological examination revealed tumor composed of sheets of highly bizarre plump endothelial cells forming anastomosing vascular channels with a high N:C ratio, marked nuclear pleomorphism, vesicular nuclei and prominent nucleoli. Brisk mitosis and numerous multinucleated giant cells were also found along with areas of extensive necrosis and blood lakes. Spleen was free of any atypia or malignancy. CD31 & CD34 were positive in neoplastic cells on IHC. The findings were suggestive of angiosarcoma. The patient is presently on follow-up and his antihypertensive medications have been stopped in view of normalization of blood pressure post-surgery. He is being planned for further follow-up imaging to assess the status of metastatic lesions.

Discussion

Adrenal angiosarcoma, first case described in 1988 in a 54-year-old male by Kareti et al., is a highly aggressive and rare malignancy [6]. The median age of presentation ranges from 34 to 85 years with a gender predilection for

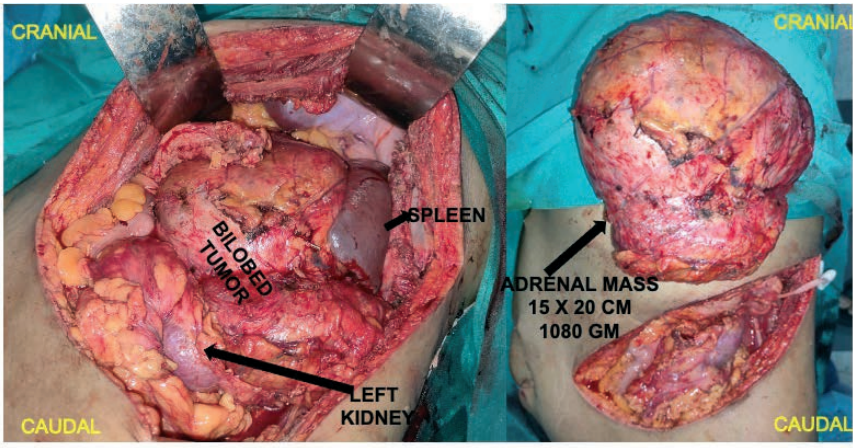

Fig. 1. Intraoperative findings of a bilobed left adrenal tumor adherent to pancreas and spleen.

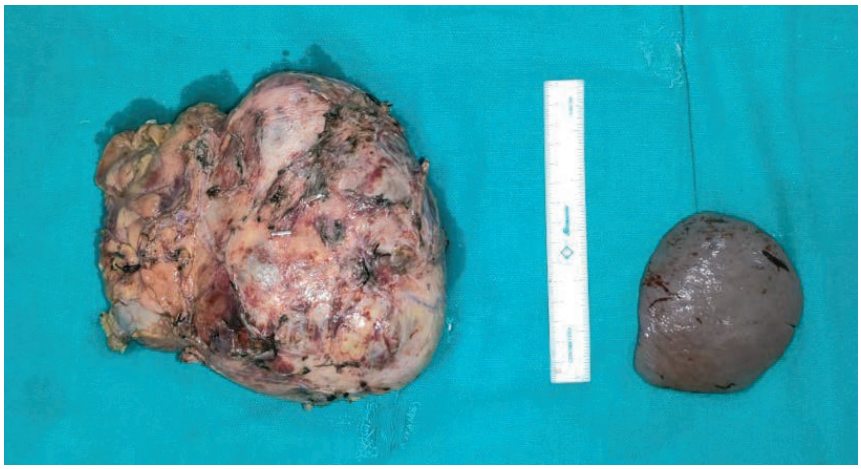

Fig. 2. Specimen of adrenal tumor along spleen.

males as reported by the countable number of cases reported in literature [7]. The symptomatology varies from typical abdominal or flank pain, palpable mass, weight loss, fatigue or weakness to being an incidental finding on imaging. Patients with systemic symptoms tend to have a poor prognosis [5,7]. The etiology of adrenal angiosarcoma is usually unknown, with few cases showing association with arsenic and vinyl chloride [3]. Hyperfunctioning tumors are yet to be reported in literature [8]. Detection of adrenal masses are on a rise due to the advances in imaging, most of them being nonfunctional and benign in contrast to this case [9]. This adrenal lesion was an incidental finding, but functional as well as malignant.

Though rare, these tumors have a high malignant potential with local infiltration as well as distant metastasis being commonly found at diagnosis [10]. This usually warrants removal of adjacent organs during surgical resection of adrenals as was with the splenectomy in our case. Being a tumor with very low incidence, there is no standardized treatment protocol described for these lesions [5]. Surgical resection usually results in a good outcome [11]. Adjuvant therapies are described for these lesions due to their aggressive nature [12].

The pathologists also have a dilemma in diagnosing primary adrenal angiosarcomas due to the non-specific histopathological characteristics with necrosis, hemorrhage and cystic components. Most adrenal angiosarcomas have a solid epithelioid pattern in contrast to other angiosarcomas with a vasoformative pattern [10, 13]. A definitive pathological diagnosis requires a wider immunohistochemistry staining in the form of CD31, CD34, FLI1 and factor VIII in addition to cytokeratins for epithelial components [14]. The tumor in our case stained positive for CD31 and CD34, thereby confirming the diagnosis.

Conclusion

This case of a rare tumor showcased the importance of multi-modality approach with Medical Oncology, Surgery, Radiology and Pathology for the challenging diagnosis as well as treatment. A standard treatment protocol needs to be developed even though the incidence of this lesion is very low. Surgical excision remains the mainstay of treatment along with adjuvant therapeutic options.

Conflict of Interests

Authors declare no conflict of interest.

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Author's Contributions

Arun Kumar M. – conceptualization, methodology, formal analysis, writing – original draft; *Nilanjan Roy* – conceptualization, methodology, formal analysis, writing – reviewing and editing, data curation, investigation; *Haris Jafri* – investigation, formal analysis, writing – reviewing and editing, data curation; *Vitesh Popli* – conceptualization, methodology, investigation, writing – reviewing and editing; *Thinley Phuntsho Roy* – methodology, data curation, investigation; *V. S. Karthik* – writing – reviewing and editing, data curation.

РІДКІСНА ПУХЛИНА - АНГІОСАРКОМА НАДНИРНИКІВ (клінічний випадок)

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Вступ. *Первинна ангіосаркома надниркових залоз є виключно рідкісною пухлиною з частотою <1% сарком м'яких тканин. У літературі описано менше 50 випадків, більшість з них були нефункціональними і супроводжувалися симптомами припухлості в животі, втрати ваги, втоми або слабкості. Лікування залишається мультимодальним із залученням хірургів та онкологів.*

Мета. *Метою був опис цього рідкісного захворювання, яке було функціональним, метастатичним і лікувалося за допомогою мультимодальної терапії.*

Методи. *Описаний випадок 47-річного чоловіка, у якого діагностували метастатичну ангіосаркому надниркових залоз, після чого було проведено хіміотерапію та згодом операцію.*

Результати. *Остаточне патогістологічне дослідження препарату та імуногістохімія виявили ангіосаркому. На даний момент пацієнт перебуває під наглядом.*

Висновки. *Незважаючи на те, що хірургічне втручання є основним напрямком лікування, роль мультимодальної терапії є наріжним каменем для лікування ангіосаркоми надниркових залоз.*

КЛЮЧОВІ СЛОВА: **ангіосаркома надниркових залоз; супраренальний; адреналектомія; судинна пухлина.**

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