A Rare Case of Adrenal Rhabdomyosarcoma

Abstract

Rhabdomyosarcoma (RMS) of the adrenal gland is a very rare entity in medical literature. Very few case reports are available in the journals. In our case report, we have shared our clinical experience of coming across such a scenario where a 43-year-old woman presented with left-sided adrenal mass, later on proved to be pleomorphic RMS. To the best of our knowledge, this is the second case of pleomorphic RMS of the adrenal gland in an adult diagnosed by light microscopy and immunohistochemical stains.

Keywords: *A rare experience, adrenal gland, rhabdomyosarcoma*

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Introduction

Rhabdomyosarcoma (RMS) is a malignant soft tissue sarcoma that is believed to develop from primitive totipotent embryonic mesenchyme. RMS is a highly aggressive tumor with a tendency for advanced and disseminated disease early in its course. The condition is the most common soft tissue sarcoma in children. However, RMS in adults is an uncommon tumor that arises mainly in the large skeletal muscles.[1-4] Pleomorphic RMS was first described by Stout in 1946.[5] More recent studies have reported that pleomorphic RMS is rare and occurs predominantly in adults. It can also be found attached to muscle tissue, wrapped around intestines, or in any anatomic location. It mostly occurs in areas naturally lacking in skeletal muscle, such as the head, neck, and genitourinary tract.

Case Report

A 43-year-old lady presented in our outpatient department with mild left lower abdominal pain and discomfort for the last 6 months. Clinical examination revealed a painless retroperitoneal mass measuring 10 cm × 12 cm, occupying left hypochondrial, lumbar, umbilical quadrants of abdomen. The patient was neither hypertensive nor diabetic. Other physical examination was unremarkable. Patient had no history suggestive of diaphoresis, sweating, paroxysomal hypertension, and headache. Patient had no history of abdominal complains in the form of hematemesis, malena, vomiting. Computed tomography of whole abdomen showed a large well-defined heterogenous inhomogenously enhancing retroperitoneal space occupying lesion (SOL) measuring 14.8 cm × 10.5 cm [Figure 1]. The lesion has displaced bowel loops, spleen and pancreatic tail anteriorly, and left kidney inferiorly. Left adrenal gland is not seen separately. Fat planes adjacent to the lesion are intact. Right adrenal is normal.

Ultrasonography-guided fine-needle aspiration biopsy from the retroperitoneal SOL was suggestive of adrenal cortical carcinoma. Value of biochemical 24 h urinary vanillylmandelic acid was normal. After exploratory laparotomy, we offered her left-sided adrenalectomy. Intraoperative period was uneventful. Postoperative histopathological analysis revealed that it was either adrenal cortical carcinoma or pheochromocytoma. On immunohistochemistry, it was favorable with the diagnosis of pleomorphic RMS as it was strongly positive to vimentin, desmin, myogenin, CD56 [Figure 2]. No expression of smooth muscle actin (SMA), synaptophysin (SYN), or S-100 protein was identified in the tumor tissue.

Discussion

In the current World Health Organization Classification of Soft Tissue and Bone Neoplasms, RMS is divided into three distinct subtypes, embryonic, alveolar, and pleomorphic.[6] RMS is a rare disease of the

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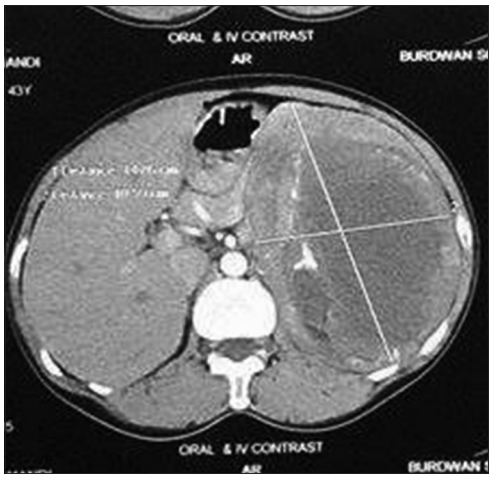

Figure 1: Computed Tomography (CT abdomen) - Shows A large well defined heterogenous inhomogenously enhancing retroperitoneal SOL measuring (14.8x10.5) cm

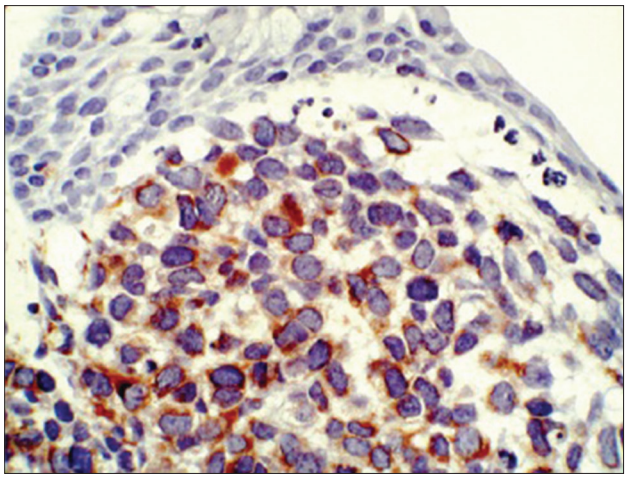

Figure 2: Immunohistochemistry-positive for vimentin

adrenal gland neoplasm, which predominantly occurs in adults. Charytonowicz *et al*.[7] suggest that RMS may arise from nonmuscle cells, including mesenchymal stem cells. Theoretically, RMS may affect any body part including the adrenal glands as shown in the present case. To date, only two RMS cases in the adrenal region have been described in English literature. Yi *et al*.[8] reported a case of alveolar RMS in the right adrenal region of a pediatric patient with a characteristic history of hypertension and fever. Katayama *et al*.[9] reported a case of RMS in the adrenal region of an elderly hypertensive patient. However, pleomorphic RMS of the adrenal gland in an adult has not been previously reported. In the present case, light microscopic examination revealed a malignant pleomorphic mesenchymal neoplasm, characterized mainly by the proliferation of atypical spindle cells and few epithelioid cells. Immunohistochemistry revealed positive staining for MyoD1, desmin, vimentin, and CD56. By contrast, no expression of SMA, SYN, or S-100 protein was identified in tumor tissue. A diagnosis of pleomorphic RMS was confirmed according to the clinical and pathological findings.

Conclusion

The present study described a rare case of pleomorphic RMS in the left adrenal region based on the immunohistochemistry results. Due to the small number of described cases of adrenal gland RMS, inadequate information is available for evaluating the treatment procedure and the final prognosis of the patient. An accumulation of such cases and an improved understanding of the molecular biology driving RMS tumor behavior are required for further evaluation and research to identify the histogenesis of the condition. Primary pleomorphic RMS of the adrenal gland in an adult is a rare condition. To the best of our knowledge, this is the second case of pleomorphic RMS of the adrenal gland in an adult diagnosed by immunohistochemical staining.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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