EWING SARCOMA OF ADRENAL GLAND CAUSING CUSHING’S SYNDROME; AN EXCEPTIONALLY RARE TUMOR

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ABSTRACT

Ewing’s sarcoma (EWS) or peripheral neuroectodermal tumors (PNET) are rare tumors. These mostly arise in the diaphyses of long bones but can also occur in various extra skeletal tissues and carry poor prognoses. Modality treatment of surgery and chemoradiation is needed to improve survival. EWS of adrenal gland is very rare and the associated Cushing’s syndrome is exceptional. Here in, we present a case of EWS adrenal gland with associated Cushing’s syndrome.

Key Words: Ewing’s sarcoma, Peripheral Neuroectodermal Tumor, Adrenal gland, Cushing’s syndrome.

INTRODUCTION

Ewing’s sarcoma (EWS) was first described by James Ewing in 1921. Similar tumors have now been grouped together, called as Ewing’s sarcoma family of tumors (ESFT). These are rare malignancies and consist of Ewing’s sarcoma of bone, extra skeletal Ewing’s, Primitive neuroectodermal tumor (PNET) and Askin’s tumor. By and large, males are more often affected than females by ratio of 1.6:1. Ewing sarcoma/peripheral neuroectodermal tumors (EWS/PNET) are usually found in 70% of the cases involving the diaphyses of long bones. But the flat bones, chest wall and extra skeletal soft tissues or occasionally, the solid organs are also involved. The extra skeletal EWS has been reported in various locations of the body, including scalp, central nervous system (CNS), nose, lips, hands, retroperitoneum, paravertebral tissues, kidney, small intestine, pelvis, cervix, vagina and rectum etc. However, the EWS of the adrenal gland is a very unusual pathology and very limited cases of primary adrenal EWS have been reported in the literature. Whereas, EWS causing Cushing’s syndrome is even more rare and exceptional. Here in we present yet another very unusual case of EWS of the adrenal gland with Cushing’s syndrome.

CASE REPORT

Eighteen years old married female, primary gravida from a very poor family in Afghanistan had a history of progressive weight gain & swollen feet since her first conception. She then developed generalized body swelling, abdominal pain and skin rash all over her body, during 7th gestational month. She consulted the local GPs and her blood pressure (BP) was noted to be increased. She was treated for hypertension and her weight gain was attributed to her pregnancy. She then started feeling headache, decreased vision, nausea increased hair fall and dyspnea on mild exertion. She kept switching from one to the other hospital and was delivered through a cesarean section in Pakistan. Her symptoms were a little bit alleviated but continued. Her post operative course remained uneventful.

In the postpartum period, her menstrual cycle remained irregular and she experienced oligmennorrhea. While, during the work up for hypertension, she was found to have Cushingoid features like obesity, moon face, plethora, striae on the abdomen and skin rash all over her body, predominantly lower limbs. An ultrasound (US) abdomen, during the routine work up showed a complex mass in the left supra-renal location.

Her serum ACTH and Cortisol were found to be within normal limits. While the contrast enhanced CT scan abdomen and chest showed a 9 into 9 cm lobulated left adrenal mass with perinephric fluid collection. There were fibro-elastic bands in the region of lingula and cardiomegaly was also noted. The echocardiog-
Ewing sarcoma of adrenal gland causing cushing's syndrome

The patient consented to publish the details of his case. Extra skeletal neoplasm has also been observed that it is meager resources, the case could not be evaluated further. Whereas, it has also been observed that it is normal levels of Cortisol and ACTH in our patient shows a 12 year old girl, having EWS of tibia. The overwhelming majority (21/32) were female, showing the EWS of adrenal to be more common in females in contrast to the gender distribution and general trend of EWS tumors, EWS of the adrenal is an extremely rare group of malignant tumors, arising in the nervous system, not exhibiting specific features which make them difficult to classify. Of all these tumors, EWS of the adrenal is an extremely rare and exceptional occurrence. Guo H and associates could gather only 32 cases of EWS of adrenal gland from the literature till 2017, including one of their own. They tabulated the gross details of all these patients. But none of them was found to have an associated Cushing’s syndrome. And of all those patients, the overwhelming majority (21/32) were female, showing the EWS of adrenal to be more common in females in contrast to the gender distribution and general trend of other EWS tumors, being more common in males.

After a thorough search and to the best of our knowledge, we could find only two cases of EWS with associated Cushing’s syndrome in the literature; one in a 9 year old girl with adrenal EWS and another in a 12 year old girl, having EWS of tibia. Moreover, the normal levels of Cortisol and ACTH in our patient shows that EWS of adrenal may secrete some other unusual peptides causing Cushing’s syndrome. While the exact cause of that needed to be found out but due to the meager resources, the case could not be evaluated further. Whereas, Technetium-99 scintigraphy is a sensitive method to detect the skeletal metastases. Moreover, Fluoro-deoxy glucose – Positron Emission tomography (FDG-PET) is also considered to be very useful aid in staging and restaging with very high accuracy.

EWS/PNET are aggressive tumors, exhibit early metastatic course and carry poor prognosis. In addition to the local lymph node involvement, these tumors metastasize to lungs, liver and bones. The tumor site, size and metastases are the strong predictors of mortality. However, EWS/PNET and especially adrenal EWS are rare tumors with no definite treatment guidelines. But a multimodality management is usually recommended. The combination of surgery along with adjuvant chemo-radiation for the loco-regional and systemic control of the disease has been found to improve the overall survival in these patients.

CONCLUSION
The reporting of this very rare and exceptional case is important since it adds to the very few and limited cases of the EWS of adrenal gland presenting with Cushing’s syndrome. For such patients, a multimodality treatment must be offered.

CONSENT
The patient consented to publish the details of his case but did not allow us to take her pictures. This case was also presented in the clinical and pathological meeting of Khyber Teaching Hospital, Peshawar, Pakistan.

REFERENCES
Ewing sarcoma of adrenal gland causing cushing’s syndrome


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AUTHOR’S CONTRIBUTION

Following authors have made substantial contributions to the manuscript as under:

Afridi ZU: Case management, references & case writing.
Haleem A: Case management.
Khan MH: Case management & references. Ahmad R: Case management.
Rashid M: References & drafting. Aroorj S: References & drafting.
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Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.