tumor may be able to produce sufficient ACTH to raise the level in plasma.

In another way of thinking, if the ACTH producing cells was present in a very limited part of the tumor, it may be difficult to find. Anyway further investigation is required for clarification.

To the best of our knowledge, ESFT with elevated ACTH levels has been reported in two children with EWS.[11] Including our patient, these three cases suggest that some ESFTs may be associated with ACTH production. EWS/FLI-1 fusion protein may be involved, as a reported transcriptional modulator for oncogenic transformation.[12]

In summary, we report the first case of renal PNET with elevated ACTH and cortisol levels. However, the relationship between ESFT and ACTH production remains unknown. Further research is needed to clarify the pathogenesis of PNET and ACTH-producing tumors.

Conflict of interest

The authors declare no conflict of interest.

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Editorial Comment

Editorial Comment to Renal primitive neuroectodermal tumor with elevated plasma adrenocorticotropic hormone levels: A case report

I read with great interest the case report by Shimizu *et al.*[1] The authors presented an interesting renal primitive neuroectodermal tumor (PNET) case. This case is very interesting in terms of patient's symptoms and age at admission and survival. The patient was 58 years old and her symptoms were whole-body edema and weakness of lower limb muscles at the time of admission. The patient was alive without recurrence 3 years after surgery.

The PNET are part of Ewing's sarcoma family of tumors and it is usually encountered in the bone and soft tissue of young adults.[2,3] In the majority of cases, patients were metastatic at diagnosis or became metastatic after a few months confirming the aggressive nature of this tumor.

To date, more than a hundred cases of renal PNETs have been reported in the current English literature. Recently, the largest meta-analysis studied 116 cases of renal PNET.[4] The median age of the cohort was 28 years (range 20–42) with 22% of patients with an age of 15 years or less. The most frequent symptoms were pain (54% of patients), hematuria (29% of patients), and renal mass (28% of patients). One third of patients were metastatic at diagnosis and 40% of non-metastatic patients developed metastasis after radical nephrectomy. In this cohort, median overall and median disease-free survival were 26.5 and 5.0 months, respectively.

More interestingly, in Shimizu *et al.*'s case, the patient had renal PNET accompanying elevated plasma adrenocorticotropic hormone levels.[1] Ectopic adrenocorticotropin syndrome could seen as paraneoplastic syndrome and it has been attributed to ectopic adrenocorticotropin secretion, which may result in several types of tumors. However, ectopic adrenocorticotropin syndrome due to PNET is extremely uncommon. The diagnosis of ectopic adrenocorticotropin syndrome was established based on the presence of adrenocorticotropin seen on immunohistochemical staining of the tumor, the disappearance of symptoms, as well as a decrease and normalization of plasma adrenocorticotropin and cortisol levels after resection of the tumor.

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The author declares no conflict of interest.

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