Elevated serum alpha-fetoprotein in Wilms' tumor: A case report with review of literature

Sushil Dhungel*, Liu Jun Cheng, Zhong Zhi Hai

Department of Pediatric Surgery, The 1st Affiliated Hospital, Sun Yat-Sen University, Zhong Shan Er Lu -58, Guangzhou 510080, PR China

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Abstract

Wilms' tumor is the second most common primary malignant intra-abdominal tumor in childhood. It is derived from primitive metanephric blastema and characterized by histopathologic diversity. Alpha-fetoprotein (AFP) is a typical oncofetal antigen of hepatic and germ cell tumors, particularly those containing yolk sac elements. Herein, the rare case of elevated AFP level and fall following chemotherapy has been described in a child with Wilms' tumor. The subject on report is a two-year old male baby detected with a palpable mass on the right side of the abdomen. His abdominal CT examinations revealed a huge mass in the right kidney, with the possibility of nephroblastoma on the right side and shadow of possible metastases in the left lung. Laboratory tests were within normal limits, but elevated serum alpha-fetoprotein (238 ng/ml). Following this, he was given three courses of preoperative chemotherapy actinomycin D, vincristine and pirarubicin (ACTD + VCR + THP). The result was that the level of AFP decreased to 82.78 ng/ml after the first two chemotherapy courses. He was then given third course of chemotherapy, the AFP level decline to 1.87 ng/ml two days prior to right radical nephrectomy. Post-operative AFP level was 1.42 ng/ml. One postoperative course of chemotherapy too was applied. Consequently he was free of disease at the age of 4 years and 5 months with unremarkable AFP blood value level.

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1. Case report

The subject on report here is a two-year old boy with a palpable mass on the right side of the abdomen. There was no prior evidence of infection or hepatitis. He was admitted for evaluation and management to Sun YatSen University, first Affiliated Hospital. Earlier, he was treated in another hospital with the complaint of a huge abdominal mass on the right side. His CT examination (Fig. 1) showed a huge mass in the right kidney, the possibility of blastoma in the right kidney and shadow of metastases in the left lung.

He was then referred to the Zhongshan University Cancer Hospital. His laboratory tests were within normal limits, but his blood serum level of neuron-specific enolase (NSE) and alpha-fetoprotein (AFP) showed 55.6 ng/ml and 238 ng/ml, respectively. Likewise, his retroperitoneal tumor biopsy and pathological diagnosis reported mixed type of Wilms' tumor. Finally his clinical diagnosis was made as Wilms' tumor stage IV. Next, he underwent three courses of chemotherapy with actinomycin D, vincristine and pirarubicin “ACTD + VCR + THP” which ran smooth with no significant adverse reactions. Upon reviewing CT after the course of chemotherapy, the size of right kidney lesion had the size of 108 mm × 98 mm, which can be considered as post-chemotherapy shrinkage. The left upper lung enhancement was 10 mm × 11 mm upon considering metastasis with multiple small lymph nodes in bilateral supraclavicular region, bilateral axillary region, abdominal aortic region and the bilateral inguinal region. Post-chemotherapy AFP level also decreased to 82.78 ng/ml, when the same
two days earlier prior to surgery was 1.87 ng/ml. Next he underwent right radical nephrectomy. Six days later, it followed with the level of AFP decrease to 1.42 ng/ml. Alpha-fetoprotein profile of the patient is shown in Fig. 2. On the eighth postoperative day he was discharged. His histopathological lab reported an excised kidney as Wilms’ tumors (epithelial type). After two weeks of surgery he received one course of postoperative chemotherapy before being discharged from the hospital. Recently again, he underwent a surgery for pulmonary metastasis from another hospital. Four years and five months later he was declared free of the disease with unremarkable AFP blood value level.

2. Pathology

2.1. Gross specimen and histopathology

Grossly (Fig. 3), the nephrectomy specimen weighed 510 g and showed a solid spherical tumor measuring right renal size of about $14 \times 8 \times 7$ cm, and tumor about $12 \times 8 \times 7$ cm. Tumor capsule was thick, tough, solid, near palpable and had lymph nodes. The cut surface was variegated with gray white, fish flesh appearance with cystic degeneration. The renal capsule was intact. There was no involvement of the renal pelvis or vein.

Microscopically (Fig. 3), tumor-infiltration was with the destruction of the renal parenchyma. The part of the tumor cells were naive glomeruloid or in glomerular-like structure. The epithelial element comprised of short spindle and a small round, scattered in the glomerular kind/tubular-like structure. Tumor infiltration read the right adrenal below the right side of the inferior vena cava & lymph nodes. No other area identified with the tumor infiltration in ureter and lymph nodes.

Immunohistochemistry: Vimentin in (+), CK (+), WT-1 (+), part of the CD56 (+), Desmin, NSE, CgA (–), Syn (more wax block) (–). A final diagnosis of specimen was Wilms’ tumor (epithelial type).

3. Discussion

Wilms’ tumor was named after the 19th century German surgeon Carl Max Wilhelm Wilms. It is probably derived from primitive metanephric blastema. The histological appearance is characterized by marked structural diversity. Classic Wilms’ tumor is composed of three types of cells — blastemal, stromal, and epithelial, although the occurrence of all three types in the same case is uncommon [7].

Although elevated level of neuron specific enolase, carcinoembryonic antigen, and hormone tumor markers, erythropoietin and renin have been reported in WT. A very few cases of Wilms’ tumor with the elevation of alpha-fetoprotein has been reported to date. The level of elevated AFP in nephroblastoma was first reported by Roth et al. in 1984 [8]. So far to our knowledge, there are only four cases reported with elevated AFP in Wilms’ tumor with the classic type [4,8–10]. Similarly, elevated AFP has been found in teratoid Wilms’ tumor, which has a predominance of teratoid elements consisting of more than 50% of the tumor. This type of teratoid Wilms’ tumor with elevated AFP return to normal after nephrectomy was reported in teratoid WT by Ashworth et al. [5]. In this case, the source of elevated AFP was demonstrated by immunohistochemistry as the AFP containing cysts lined by enteric-type epithelium within the tumor. In this particular case, teratoid elements and enteric foci were not observed.

Patriarca et al. in 1992 had described about a case of Wilms’ tumor with elevated AFP. In that report the tumor did not show regression either on clinical or pathologic assessments, and serum AFP levels decreased upon preoperative chemotherapy, but...
returned to normal limits after nephrectomy. This report is very similar to ours, as our case showed a dimensional regression upon preoperative chemotherapy when AFP level too decreased, which later returned to normal level after nephrectomy. Similarly Kismet Erol in 2005, A. Crocoli in 2008 and V. Kesik et al. in 2010 had reported Wilms’ tumor with elevated AFP level, which later returned to normal level just after nephrectomy. Similarly Kismet Erol in 2005, A. Crocoli in 2008 and V. Kesik et al. in 2010 had reported Wilms’ tumor with elevated AFP level, which later returned to normal level just after nephrectomy. AFP values were far higher in patients with metastatic disease than in the one with thrombosis of inferior vena cava [10]. As in our case the metastasis was only found in lungs.

Like any elevated tumor marker, elevated AFP by itself is not diagnostic, but is only suggestive. Tumor markers are used primarily to monitor the results of a treatment. If levels of AFP go down after treatment, the tumor shows not growing. In the case of babies, after treatment AFP should go down faster than it would normally. A temporary increase in AFP immediately following chemotherapy may indicate that the tumor is growing, but rather that it is shrinking. Rare AFP-secreting tumor types include carcinoma in a mixed Müllerian tumor [11]. The Sertoli-Leydig cell tumor, which itself is rare, rarely secretes AFP [12]. In Wilms’ tumor AFP is rarely elevated, but when it is elevated, it may be read as a marker of disease progression or recurrence [10]. Increased serum levels of alpha-fetoprotein are sometimes found in Citrullinemia and Argininosuccinate synthetase deficiency [13].

4. Conclusion

Usually, there are no criteria for performing the routine serum AFP sampling in renal tumor due to its easy clinical diagnosis. Due to this, chance was that the past had missed a large number of elevated AFP level cases of Wilms’ tumor. After the diagnosis of this case, we sent the investigation report of all new patients with Wilms’ tumor. Here then no other such case was reported existing with the elevated AFP level. We rather recommend now sending AFP level in all cases with renal tumor as the first line of investigation.

References