Testicular yolk sac tumor in an eight-month old child: A case report

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ABSTRACT
Germ cell tumors are rare in children. This group of tumors in children is distinct from that in adults in terms of their behavior, histological nature, metastasis and the treatment needed. 85% of the yolk sac tumors in children present as clinical stage I disease as compared to 35% in adults. AFP can be used as a reliable tumor marker as the levels are increased in more than 90% of yolk sac tumors. In children inguinal orchidectomy followed by stringent surveillance for two years is the preferred treatment option. Combination chemotherapy may be reserved for metastatic disease or recurrence. Retroperitoneal Lymph Node Dissection (RPLND) is preferable in adults. In this case report we describe the clinical presentation, imaging findings, diagnosis and up-to-date management of yolk sac tumor in an eight-month old child.

Key words: germ cell tumor, yolk sac tumor, alpha fetoprotein, chemotherapy, RPLND

INTRODUCTION

Testicular tumors are rare in children, but highly treatable and usually curable. In children testicular tumors account for approximately 1 to 2% of all tumors, with an incidence of 0.05-2 per 100,000 children. Among testicular tumors germ cell tumors have a bimodal age distribution. One peak occurs in the first two years of life and the second in young adults 15-35 years of age. Pediatric testicular tumors are dramatically different from adult neoplasms. Germ cell tumors account for 60% testicular tumors in children, but 95% of testicular tumors in adults. Adult germ cell tumors like seminoma and embryonal carcinoma are rare in children. Teratoma which are benign in children are often malignant in adults.

This is the first case of testicular cancer, yolk sac tumor, in an eight-month old child in the author's 27years of experience in urology.

CASE REPORT

A mother brought an eight-month old child to the Urology OPD with a painless right testicular swelling, which was noticed one week previously. This was the first clinic that the child had attended. On examination it was 6x5cm in size, non-tender, irregular in shape and firm in consistency. There was a family history of cancer. The uncle of the child had died at the age of 18 years because of leukemia a few years previously.

Color Doppler ultrasound examination revealed an enlarged right testis measuring 4x3x2.5cm with estimated volume of 16.3cc. It showed multiple hypoechoiccystic foci of different sizes and appeared hypervascular on color Doppler evaluation. Hypervascularity was also noticed within the right spermatic cord. A mild amount of free fluid was detected in the right scrotal compartment. As the normal size of the left testicle for this age is about 1.7x1.1x0.6cm with an estimated volume of 0.56cc. the clinical diagnosis made was enlarged right testicle, possibly neoplastic in nature.

MRI of the upper abdomen, pelvis and male perineum revealed a diffusely enlarged right testis measuring 4.6x2.8x4.4cm, displaying heterogenous high signal in T2 and fat suppressed images with central necrosis and appreciating heterogenous

yet significant post contrast enhancement, a pattern that is indicative of a neoplastic process. The ipsilateral epididymis was diffusely enlarged with significant post contrast enhancement. Mildly thickened and enhancing ipsilateral spermatic cord. The right scrotal compartment was distended by the diffusely infiltrated testis with associated minimal hydrocele and diffuse thickening and enhancement of the surrounding skin. The left testis was displaced in an anterior and superior direction by the mass effect and noted in the left scrotal neck and measuring 16x8mm in diameter. A few bilateral inguinal LN with no suspicious criteria were observed. Additionally, liver, spleen, gall bladder, pancreas, kidneys, major vessels and urinary bladder were found to be normal. No detectable masses or lymph nodes were seen in the abdomen. Both lung fields and C/P angles were clear on chest X-ray.

The values of tumor markers Alpha Fetoprotein (AFP) and betaHCG are shown in Table 1.

Table 1.Values of tumor markers

Tumor Marker	Observed Value	Biological Reference Range
AFP	29618ng/ml	0.2-9.0 ng/ml
Beta HCG	0.500miu/ml	Less than 5 miu/ml

The patient underwent unilateral orchidectomy through inguinal approach. The right testis was removed together with all its coverings with maximum cord length up to the deep inguinal ring. Spermatic veins were observed to be dilated and tortuous. No fixity to skin was seen. The patient was discharged the next day and stitches removed after eight days. The post-operative period was uneventful.

Gross examination of the specimen submitted for histopathology revealed a well circumscribed gray-white neoplasm 4x4x3cm, with areas of necrosis and cystic change, almost totally replacing testicular parenchyma (Figure 1).

H&E stained sections of the tumor showed cuboidal tumor cells arranged mostly in a reticular-microcystic pattern. The cells exhibited pleomorphism, hyperchromatisia and frequent mitosis with occasional intracellular and extracellular eosinophilic hyaline globules. Glomeruloid papillary structures composed of tumor cells covering a central fibrovascular core within a cystic space lined by a single layer of flat/cuboidal cells (Schiller-Duval Bodies) were seen in some areas (Figure 2). Solid and microcystic patterns were also observed. No seminomatous, choriocarcinomatous or teratomatous elements were identified. A few compressed seminiferous tubules were present at the periphery of the tumor. Sections from the epididymis, spermatic cord and rete testis were free of tumor.

Immuno histochemical staining was negative for CD30 and focally positive for AFP. The findings were consistent with pure yolk sac tumor.

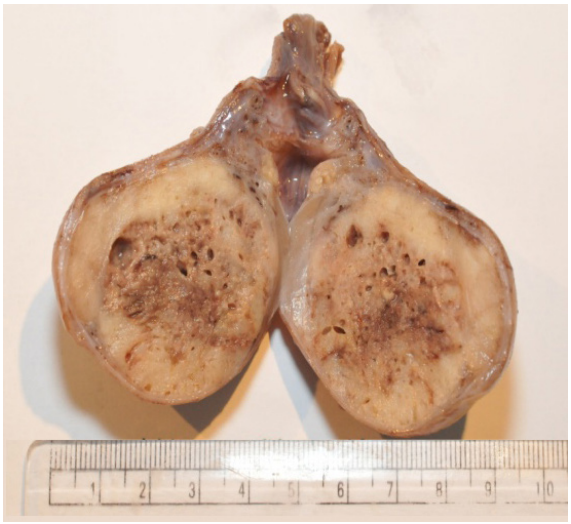

Figure 1.Cut section of the testicular tumor revealing a circumscribed solid gray-tan neoplasm with necrosis and cystic change.

The patient is under stringent follow-up. AFP analysis done 17days after surgery had dropped significantly to 636 ng/ml from its 29618 ng/ml before surgery. He was referred to the oncologist for further management and follow up.

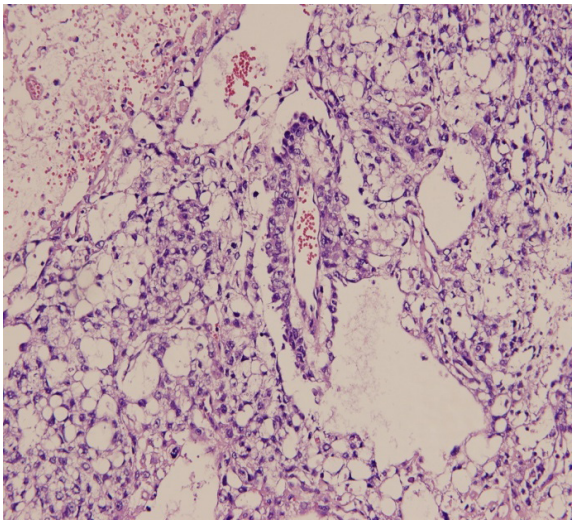

Figure 2. Schiller–Duval body (arrow) characterized by tumor cells forming glomeruloid papillary structure is pathognomonic of yolk sac tumor. (H&E x20)

DISCUSSION

Germ cell tumors are most frequently found in the gonads. Only 2-5% of them arise in extragonadal regions such as mediastinum, retroperitoneum, pineal gland and sacral areas. Yolksac tumor of penile shaft and urachus has also been reported[2]. Two types of germ cell tumors are seminomas and non-seminomas. Seminomas are extremely rare in prepubertal children. Among non-seminomatous germ cell tumors the most common are teratomas and yolk sac tumors which account for about 62% and 26% of testes tumor in infants and toddlers respectively[3]. Teratomas are described in some series as constituting 50% of prepubertal testicular tumors. However these are reported less because of their benign nature[5]. In tumor registries, yolk sac tumors are mentioned more commonly than teratomas[1].

Different risk factors have been described for testicular cancer in the early age group. They include pre-natal exposure to high estrogen levels during intrauterine life, gonadal dysgenesis, exposure of mother to chemical carcinogens and smoking, trauma, testicular atrophy due to torsion, and orchitis.

Family history of cancer is an important risk factor in the early age groups. In the case of this child the uncle had died of leukemia at around 18years of age. However no other causal relationship has been established.

Three clinical stages for the determination of extension of the tumor have been described. Stage I is where the tumor is confined to the testis. Invasion of epididymis tunica albuginea, spermatic cord or scrotum does not change tumor stage but increases the risk of nodal involvement and the risk of recurrence. In stage II the tumor has retroperitoneal lymph node metastasis. Stage III is characterized by supraclavicular lymph nodes, visceral involvement or persistently elevated tumor marker values[4].

The vast majority, 85%, of yolk sac tumors in children present as clinical stage I disease compared to 35% in adults[1]. AFP can be used as a reliable tumor marker as levels are increased in more than 90% of yolk sac tumors. Therefore patients can be safely managed with observation after orchidectomy. Chemotherapy is reserved for recurrence. In our case, the level of tumor marker AFP was 2,9618ng/ml before surgery. MRI revealed no lymph node involvement retroperitonealy. X-ray chest was also clear. Our patient was in stage IC-T1 No Mo S3.S3 means AFP more than 10,000ng/ml. Tumor marker AFP dropped significantly to 636ng/ml on 17th post-operative day.

The nature of yolk sac tumor is different in children from that in post-pubertal adults. In young children yolk sac tumors are predominantly pure in histology and are not usually mixed with other histological types as may be encountered in adults. Histopathology and immunohistochemistry revealed that our patient had pure yolk sac tumor.

Yolk sac tumors in children metastasize through the hematogenous route in more than half of the cases in comparison to adults where it is only 4-6%[5]. This fact changes the treatment modality. It indicates that RPLND would not be adequate treatment in children and would result in complications like chylous ascities, post-operative bowel obstruction, wound infection and subsequently ejaculatory dysfunction.

The German Society of Pediatric Oncology reported that 15% of patients had recurrence with stage I yolk sac

tumor after radical inguinal orchidectomy and observation alone. These patients were cured successfully by combination chemotherapy, with 100% survival. Even in the patients with metastatic disease that they treated, four out of five had remission with chemotherapy[5-8].

Keeping the above in view we recommended observation after inguinal orchidectomy and stringent follow-up for two years for our patient. The latter includes tumor marker AFP, x-ray chest every two months and MRI every 3-4 months. If there is no evidence of recurrence after the second year, surveillance using all these methods can be less frequent since the risk is highest in the first two years.

CONCLUSION

Pre-pubertal testicular tumors are rare but result in great anxiety and burden on the family when they do occur. Testicular germ cell tumors in infants are distinct from those in adults. Despite good prognosis, the parents have many reservations regarding the treatment and its effects on the child's growth and particularly on future fertility.

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