

Case Report

Testicular embryonal rhabdomyosarcoma: A case report

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Rhabdomyosarcoma is one of the most frequent soft tissue sarcomas in children. It is found mostly in children, primarily infants, toddlers, and pre-school pupils. However, pure testicular rhabdomyosarcoma is a very rare tumor and few cases have been reported in literature. The tumor usually presents as a painless testicular enlargement with early dissemination via the blood stream and lymphatics. The origin of this tumor is presumed to be from overgrowth of a sarcomatous area of the teratoma. Here, we present a case of testicular rhabdomyosarcoma in a 15-year-old student with a fatal outcome during chemotherapy. This is actually the first case of pure testicular rhabdomyosarcoma diagnosed in this centre after over 10 years.

Key words: Rhabdomyosarcoma, testicular, orchidectomy, chemotherapy.

CASE PRESENTATION

The index case is a 15-year-old male student who presented with a two months history of right sided testicular swelling that has rapidly increased in size since onset. There was an associated dragging sensation, and no evidence of urinary tract infection. Past medical and surgical histories were also nil of note. He is the first of two sons. He lost his mother about a year earlier to invasive ductal carcinoma of the right breast. There has not been a previous history of cancer recorded in the extended family before now. On physical examination, the mass measured 16 by 6 cm and was firm to hard in consistency; it was irregular in shape and non tender. The mass appeared to be continuous with the right testis, mobile and not fixed to the scrotal skin. Other systems examined were normal. Preliminary fine needle aspiration done at the referring hospital suggested a small round blue cell tumor. On admission at the Jos University Teaching Hospital, chest x-ray, abdominal ultrasound, full blood count, urinalysis, serum urea, electrolyte and creatinine, alpha fetoprotein and - human chorionic gonadotropin were all normal. No intra-abdominal lymphnode enlargement was visualized at ultrasonography.

However, testicular ultrasonography confirmed a testicular mass that was continuous with the right testis. The left testis was normal. The boy later had a right sided high orchidectomy.

GROSS AND MICROSCOPIC FINDINGS

At the histopathology laboratory, the tissue which was preserved in 10% formal saline consists of a firm encapsulated grayish white mass of 16 × 10 × 6 cm and weighing 80 gm. Cut section showed a solid grayish mass of 14 cm diameter attached to the testis. Histology showed sheets of rhabdomyoblasts having round hyperchromatic nucleus and brightly eosinophilic cytoplasm, and several strap cells disposed in a myxoid background (Figures 1a and b). The resection margin was free of tumour cells. A diagnosis of embryonal rhabdomyosarcoma was made.

Based on this report he was referred to the surgical oncologist for chemotherapy. All earlier investigations were repeated by the oncologist before the chemotherapy to form a baseline and to rule out any evidence of occult metastasis with additional investigations like electrocardiogram. They all turned out to be normal. Exactly a month after presentation, he was commenced on a VAC cytotoxic regimen (vincristin, adriamycin and

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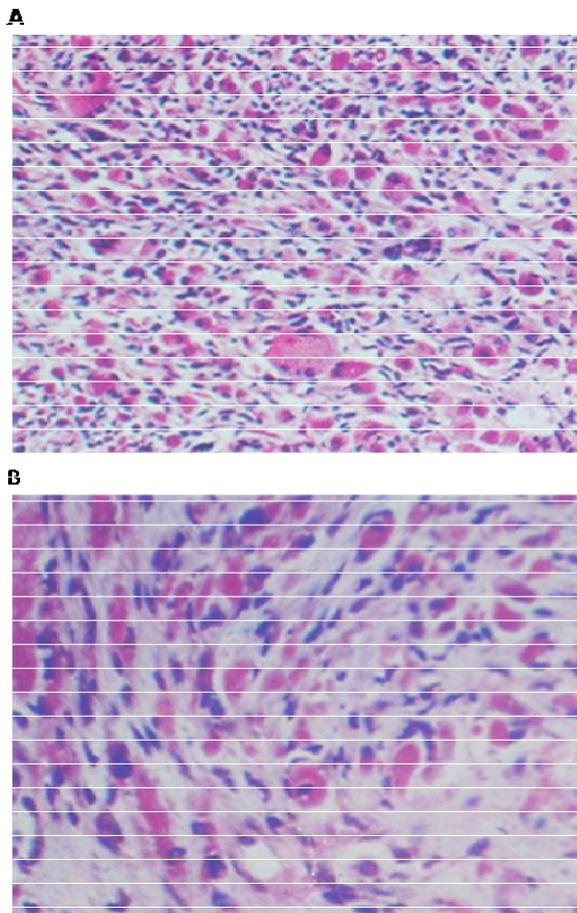


Figure 1. Embryonal rhabdomyosarcoma showing; (a) several rhabdomyoblasts having brightly eosinophilic cytoplasm and ovoid to spindle nucleus on a myxoid background, 20× objective and; (b) strap cells left half of slide, 40× objective. H & E stains.

cyclophosphamide) as an out-patient to be cycled every three weeks through 6 courses at the first instance. Routine investigations were to be done before each cycle of cytotoxics. He went through the first 6 courses without any sequelae after which he was thoroughly reviewed. He was found to be stable; hence the oncologist recommended 3 more cycles. However, after the eighth cycle, the boy all of a sudden developed severe anaemia with associated vomiting and weakness. He was admitted and transfused five pints of whole blood but later died within two days on admission. This was approximately eleven months after presentation.

DISCUSSION

Testicular tumors have been reported by Lawrence et al. (1965), as occurring in between 0.58 and 2.09% of all male tumors, and that 1 to 2% of testicular tumors is sarcomatous and only a small percentage of these are

rhabdomyosarcomas. The American College of Chest Physicians (2005) reported that testicular tumors constitute a very small percentage of all malignant tumors in men, and account for 11.4% of deaths from cancer in males between 20 - 35 years old. Trauma, cryptorchidism, and exogenous maternal estrogen (*in utero*) have all been associated with its development. Genetic mutations affecting chromosomes 1, 3 and 13 have been found in the rhabdomyosarcomas in a study by David et al. (2006). The most common presentation is pain, swelling or hardness of the testis. Alexander (1968) reported that the age of the patients varied from 3 months to 67 years. Chung et al. (2007) in Korea reported a case of pure testicular embryonal rhabdomyosarcoma in a 9-month-old baby. Rhabdomyosarcoma is regarded as a highly malignant tumor with frequent recurrence and dissemination via the blood stream and lymphatics. A few patients may already have signs or symptoms of metastatic disease such as back pain, cough and dyspnea (indicating pulmonary metastasis), nausea and vomiting, bone pain, or central nervous system manifestations, according to the American College of Chest Physicians (2005). Diagnosis is made by CT scans, serum tumor markers and surgical biopsy. Orchidectomy followed by chemotherapy and/or radiation is the treatment of choice for non-seminomatous tumors. The American College of Chest Physicians (2005) reported that recurrence may occur within 2 years so intensive surveillance and follow up is necessary. Since these tumors metastasize via lymphatics as well as the blood stream, lymphangiography and dissection of retroperitoneal nodes may be of benefit. Kumar et al. (1987) reported that little evidence is found in the literature to suggest that any treatment other than surgical removal is of value in the treatment of rhabdomyosarcoma; radiation and chemotherapy appear to have little effect. Our case is a unilateral circumscribed mass in the right testicle. Abdominal ultrasound did not show evidence of associated retroperitoneal adenopathy, and histology did not reveal tumour cells in the resection margin. However, the fear of occult metastasis resulted to the administration of the VAC regimen which obviously precipitated a fatal complication. Except in children where prognosis is poor, long term survival has been reported by Alexander et al. (1968). In our environment, the common drugs used for most of the childhood round blue cell tumours are the members of the VAC regimen, all of which are known marrow depressants. This particular mortality was purely drug related untoward effect.

Conclusion

Pure testicular rhabdomyosarcoma is rare. Managing it relies mainly on early detection and orchidectomy. The application of chemotherapy and or radiotherapy demands thorough pre, intra/inter course and post therapy evaluations.

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