Case Article

Testicular Embryonic Rhabdomyosarcoma, Case report with brief literature review

Ahmed M. Adam[1], Mohammed MA.M Ibnouf[2], Ishraga A. Faraga Allah[3]

Background:

Rhabdomyosarcoma (RMS) is a malignant solid tumour arising from mesenchymal tissues which normally differentiate to form striated muscle. It can occur in a wide variety of sites. It is one of the most frequently occurring soft tissue sarcomas and commonest in children under age of 15 years[1]. Approximately 350 new cases are diagnosed in the United States each year[2].

Seven to 10% of primary genitourinary tumours are located in the para-testicular region. Scrotal rhabdomyosarcomas originating from paratesticular tissue are most frequently seen in childhood and young adults[3]. Paratesticular rhabdo-myosarcomas are very rare, comprising only 1: 2,000 tumours of the male genital system[4]. It constitutes 7% of all rhabdomysarcomas[5].The disease may be subdivided into embryonal (which accounts for about 60%), pleomorphic, alveolar and botryoidal types. Embryonal RMS is most commonly found in head and neck, genitourinary and retroperitoneal sites[6]. The tumour is slightly more common in boys and males (11.8 per million) than in girls and females (10.3 per million)[2]. The age incidence varies from 21 months (Sabrazes et al, 1923) to 67 years (Prince, 1942)[3]. However, some had reported racial and gender differences in the incidence of RMS[7].

Here we report an adult male of paratesticular embryonal RMS. He was lost trace for a while till he present with advanced metastases to the retroperitonium and a multidisciplinary management was held later.

Keywords: Paratesticular tumours, Retroperitoneal metastasis.

A 23-year old single male from South Sudan presented to the casualty of general surgery in Albangadeed Hospital in Khartoum North city in July 2009, with left testicular pain and swelling for two and a half months. The swelling increased gradually in size. Other systems revealed insignificant history. He had no family, drug or social history relevant with his complaint. On examination the patient was stable with good general health but rather anxious. All systems were normal except his left hemi-scrotum that contained an obvious swelling. The right testis was normal. There was no local or regional lymphadenopathy. Ultrasound showed that the left testicle was grossly enlarged with heterogeneous echogenicity, lobulated structure, epididymal thickening and turbid vaginal hydrocele. The right testis and epididymis were normal but with a small right hydrocele. The tumour markers (α feto protein, β HCG,) were within normal range but LDH was two folds high. Ultra sound scan and CT scan of the abdomen reported urinary bladder wall thickening and calcification consistent with urinary bilharziasis. Urine was positive for schistosomiasis and chest radiography was clear. Trans-inguinal orchidectomy was done and histopathology revealed an irregular mass of soft tissue measuring 12×11×7cm, with a five cm segment of spermatic cord. Microscopically, section displayed proliferation of sheets of spindle cells with eccentric nuclei and acidophilic cytoplasm, with scattered strap cells, multinucleated giant cells and tumour cells with smooth muscle actin was positive. Hence, the pathological diagnosis was embryonal rhabdomyosarcoma. The spermatic cord cells were free of malignancy.

1. Assistant professor of surgery Omdurman Islamic University .

2. Prof. of Surgery, Omdurman Islamic University

3. Consultant histopathologist.

Seven days later the patient was discharged with appointment to Radio Isotope Centre- Khartoum (RICK). Thereafter, he disappeared to come back six months later i.e. on January 2010 complaining of abdominal pain and swelling for three weeks. On examination there was abdominal tenderness and a palpable firm mass about 14×20 cm. Ultra sound scan showed moderate ascites and multiple, well defined, rounded iso-echoiec retroperitoneal masses, anterior to inferior vena cava and aorta. These masses were compressing the left renal pelvis causing moderate hydronephrosis and displacing the left renal vein anteriorly. Computed tomography with I.V contrast showed a mass measuring 9.3×6.9 cm in the left para-aortic area encasing the left renal pelvis and vessels with marked central necrosis. The left kidney showed faint function and is hydronephrotic. Liver, right kidney, pancreas and spleen were normal (figure 1).

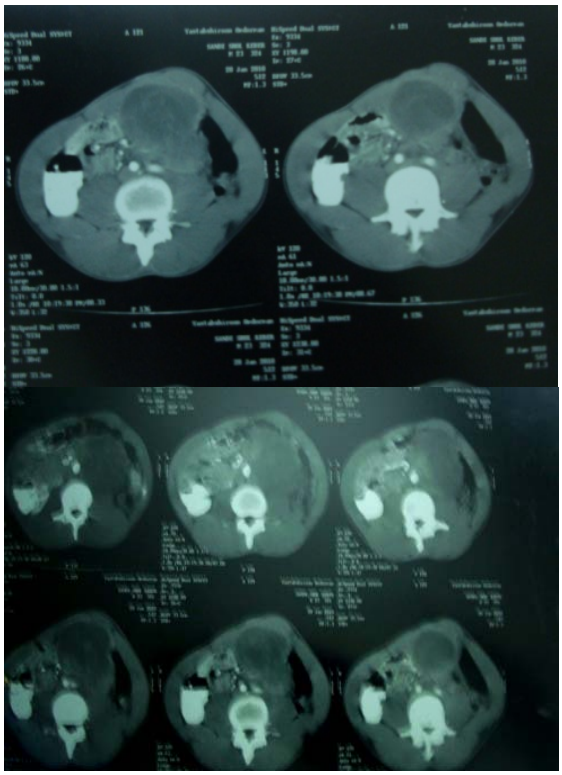

Figure (1) Computed tomography scan showing a mass in the left Para-aortic area encasing the left renal pelvis and vessels with marked central necrosis.

Exploratory laparotomy performed on 16th of February 2010 through a midline incision, debulking of the tumour was done to relieve the compression on the left renal vessels. The patient received four pints of blood and recovered smoothly from anaesthesia. The resected specimen was taken for histopathology. Histopathology reported focal necrosis, haemorrhage and fascicles of round to spindle cells with hyperchromatic nuclei and scattered strap cells. Immunohistochemistry proved presence of Desmin and Myoganin (figure 2).

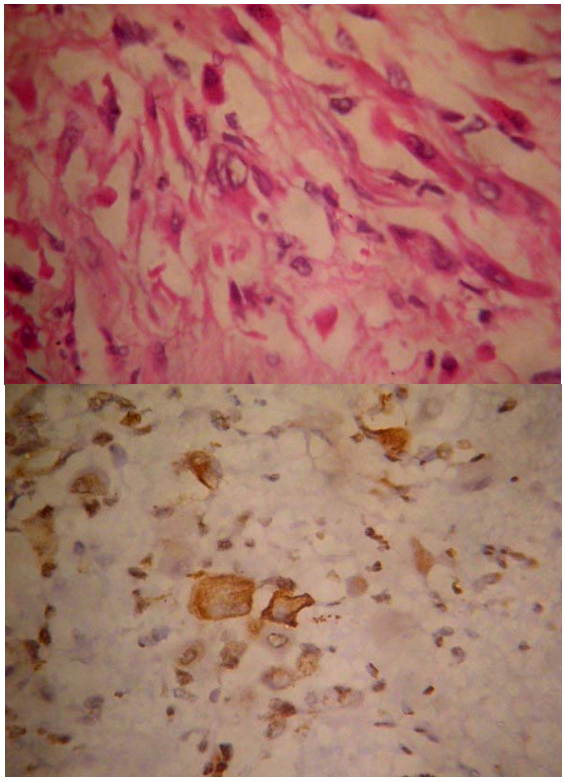

Figure (2) Tumour section, H&E stain and Desmin stain were positive ×40.

Fifteen days later the patient referred to the oncology department in RICK. A combination of Ifosfamide 180mg/m^2, Mesna 360mg/m^2 and Etoposide 100mg/m^2 was started and six cycles were completed. The follow-up CT scans showed multiple small pelvic deposits denoting no response to Ifosfamide.

Again the patient didn't come for follow-up, but returned on June 2011 with a huge abdominal mass associated with a dull

aching pain. He was referred back to the department of oncology. CT scan to the abdomen was done and the mass was again encasing the aorta and the inferior vena cava. Laparotomy was done but unfortunately the mass was not resectable.

Discussion:

Rhabdomyosarcoma may occur as a tumour in striated muscle or outside the normal sites of skeletal muscle. The latter type typically arises in the urogenital system where it is believed to be derived from embryonic rests[3]. Our case resembles this type. However, striped muscle and rhabdomyoblasts are not uncommon findings in testicular teratomas. Unilateral development of the rhabdomyoblastic element to produce a pure testicular rhabdomyosarcoma is much rarer[3]. The tumour cells may be round or oval, "strap like" with two or more nuclei arranged in tandem (hyperchromatosis) as in our case, or "racquet" shaped with a long tapering cytoplasmic prolongation (spindle) and the nucleus situated in the rounded head. Giant cells are fairly frequently observed and the vacuoles that contain glycogen are not uncommon. The cytoplasm of the rhabdomyoblast is typically eosinophilic that was not in our case which was acidophilic, and may contain cross-striations or longitudinal myofibrils. Alveolar and myxomatous areas are often observed and the histological variations have been classified as pleomorphic, alveolar, embryonal and botyroid, the last typically occurring in submucosal sites[3].

Although these tumours may arise anywhere in the body, certain distinctive clusters of features emerge regarding age at diagnosis, site of primary tumour, and histology. For example, head and neck tumours are most common in children younger than eight years of age. Especially if arising in the orbit, they are usually of the embryonal variety. On the other hand, extremity tumours are seen more commonly in adolescents and are more frequently of the alveolar subtype. A unique form of RMS arising from the bladder or vagina (submucosal); the botyroid variant (so named because of its resemblance to a protruding cluster of grapes) is seen almost exclusively in infants[7].

The clinical presentation tends to include a short history of a few weeks of painless swelling of the scrotum. The tumour has no preference for the left or right testis or for any particular race[8,9]. It has generally been accepted that sarcomas tend to recur locally or spread by the blood stream, following invasion of the thin sinusoidal channels within them. However, as more of these cases are reported there is an increasing awareness of lymphatic spread. Many reports refer to RMS as retroperitoneal masses[10,11]. Limited post-mortem findings indicated retroperitoneal metastases, invasion of the pancreas and extension to the porta hepatis without liver involvement without evidence of lung secondaries. Such findings suggest lymphatic spread without vascular spread. Considering prognosis of 11 cases[12], Gowing and Morgan did not comment on the mode of spread. However they stated that one case had massive intra-abdominal lymph-node involvement within one month of removal of the primary growth. This is the same with our patient but within six months. It must not be concluded from the foregoing statements that lymphatic spread is common method of spread of RMS. Yet, clinical findings such as infiltrated sternal and iliac marrow in the absence of palpable lymph nodes strongly suggest vascular dissemination[10]. Severe cough and right chest pain, with X-ray evidence of massive metastases were the first evidence of tumour spread as reported by Hoffman and Baird[13], again suggesting primary vascular spread, though this may of course have followed lymphatic involvement to thoracic duct level [4]. Unlike our case, one third of patients have metastases at the time of presentation.

The management of this disease has dramatically changed over the years. Today the emphasis is on a multidisciplinary approach. Before 1960, localized RMS treated with either surgery or radiation

therapy was less than 30%. However, the survival rate has risen to 90% in cases treated with surgery and chemotherapy and/or radiotherapy[14.15].

References:

1. Arya K, Vij H, Vij R. Rhabdomyosarcoma of mandible: A diagnostic predicament. 2011; 15(3) : 320-325
2. Gurney JG, Young JL, Roffers SD, et al. Soft tissue sarcomas. In: Ries LA, Smith MA, Gurney JG, et al. eds.: Cancer incidence and survival among children and adolescents: United States SEER Program 1975- 1995. Bthesda, Md: National Cancer Institute, SER Program, NIH Pub. 1999, No. 99-4649, pp 111-123.
3. Alexander F. Pure testicular rhabdomyosarcoma. British journal of cancer. Vol. XXII, No. 3: 495-501.
4. Alexander F. Intra-scrotal sarcomas. British journal of cancer, Vol. XXII, No. 3: 486-97.
5. Stewart LH, Lioe TF, Johnston SR. Thirty year review of intrascrotal rhabdomyosarcoma. Br J Urol. 1991; 68(4):418–20.
6. Kelland LR, Bingle L, Edwards S et al. High intrinsic radiosensitivity of a newly established and characterized human embryonal rhabdomyosarcoma cell line. Br. J. Cancer. I989; 59: 160-164.
7. Stiller A, Parkbin DM. International variations in the incidence of childhood soft tissue sarcomas. Pediatr Perinat Epidemiol. 1994; 8: 107-19.
8. Ferrari A, Casanova M, Massimino M, et al. The management of paratesticular rhabdomyosarcoma: a single institutional experience with 44 consecutive children. J Urol. 1998; 159(3):1031- 4.
9. Elsasser E. Tumors of the epididymis. Recent Results. Cancer Res 1977; 60: 163-75.
10. Ravich L, Lerman H, Sands A. Intrascrotal extratesticular rhabdomyosarcoma, J Urol 1964; (92): 144-7.
11. Rosas-Uribe A, Luna MA, Gene A. Guinn GA et al. Paratesticular rhabdomyosarcoma: A clinicopathologic study of seven cases. Am J Surg. 1970; 120(6): 787-91.
12. Gowing NFC, Morgan AD. Paratesticular tumours of connective tissue and muscle. Br I Urol (suppl). 1964:36:78. 535.
13. Hoffman WW. Baird SS. Soft tissue sarcoma. J.Urol.1960; 84, 376.
14. Jaffe N, Filler RM, Farber S et al. Rhabdomyosarcoma in children. Improved outlook with a multidisciplinary approach. Am J Surg 1973; 125 (4): 482-7.
15. Stevens MC, Rey A, Bouvet N et al. Treatment of nonmetastatic rhabdomyosarcoma in childhood and adolescence: third study of the International Society of Paediatric Oncology--SIOP Malignant Mesenchymal Tumor 89. J Clin Oncol. 2005;23(12):2618-28.