

PARATESTICULAR RHABDOMYOSARCOMA: A CASE REPORT

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ABSTRACT

Paratesticular rhabdomyosarcoma is a rare non germ cell tumour of scrotal sac in children and young adult/teens which can invade testis at presentation. Embryonal variant is the most common type. 40% cases can have metastasis to retroperitoneal lymph node. Diagnosis can be done on high degree of clinical suspicion coupled with biopsy and immunohistochemistry. Multimodality approach of treatment is often beneficial for patients. Here we report a case of 17-year old boy with right paratesticular solid mass. Ultrasound revealed vascular paratesticular mass separated from the testis. Right Inguinal orchiectomy was done. Histopathology revealed Rhabdomyosarcoma paratesticular. Abdominal ultrasound and CT show retroperitoneal lymph node mass. The patient was treated with chemotherapy et reoperated for lymphadenectomy but died after 4 months.

KEYWORDS: Paratesticular; Rhabdomyosarcoma; Multimodal treatment.

CASE REPORT

A 17-year-old patient, without antecedents, presented to emergency with right painless paratesticular solid mass measuring about 12 cm in its largest diameter. Abdominal examination revealed a non-tender, palpable mass in the right upper quadrant extending to below the umbilicus. Physical examination revealed no cervical, axillary or inguinal lymphadenopathy. An ultrasound revealed an expansive process characterized by intrascrotal heterogeneous tissue density. The tumor

markers α - fetoprotein, lactate dehydrogenase, and β -human chorionic gonadotropin were normal.

An inguinal orchiectomy was performed. A histological examination of the surgical specimen confirmed the diagnosis of an paratesticular rhabdomyosarcoma. Abdominal computed tomography (CT) revealed a right retroperitoneal mass measuring 13/9/8cm with a heterogeneous and solid component extending on the right side of the aorta and right ureterohydronephrosis. (figure 1).

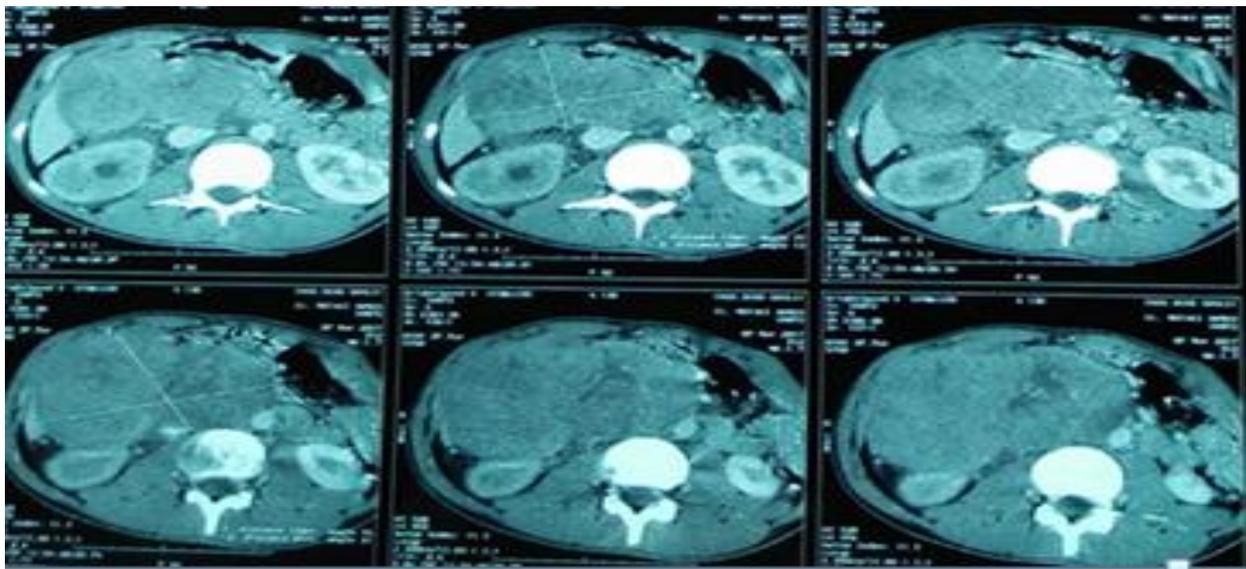


Fig. 1: Abdominal CT scan showing right retroperitoneal mit hydronephrosis.

He received multidrug chemotherapy consisting of vincristine, adriamycin, cyclophosphamide, cisplatin, and actinomycin. Follow-up CT after chemotherapy

revealed that the right para-aortic lymphadenopathy had decreased in size to 6.6×5.8 cm (figure 2).

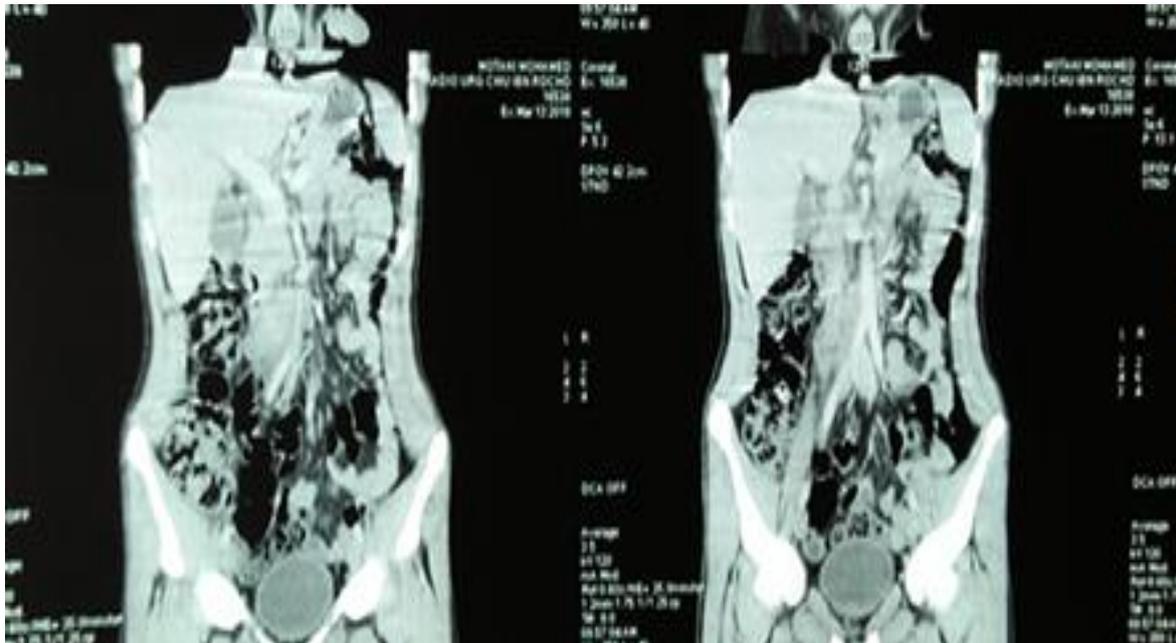


Figure 2: Abdominal CT Scanner revealed regression of the retroperitoneal mass.

We decided to do a retroperitoneal lymphadenectomy and laparotomy found a fixed mass at the inferior vena cava with no separation limit (figure3).

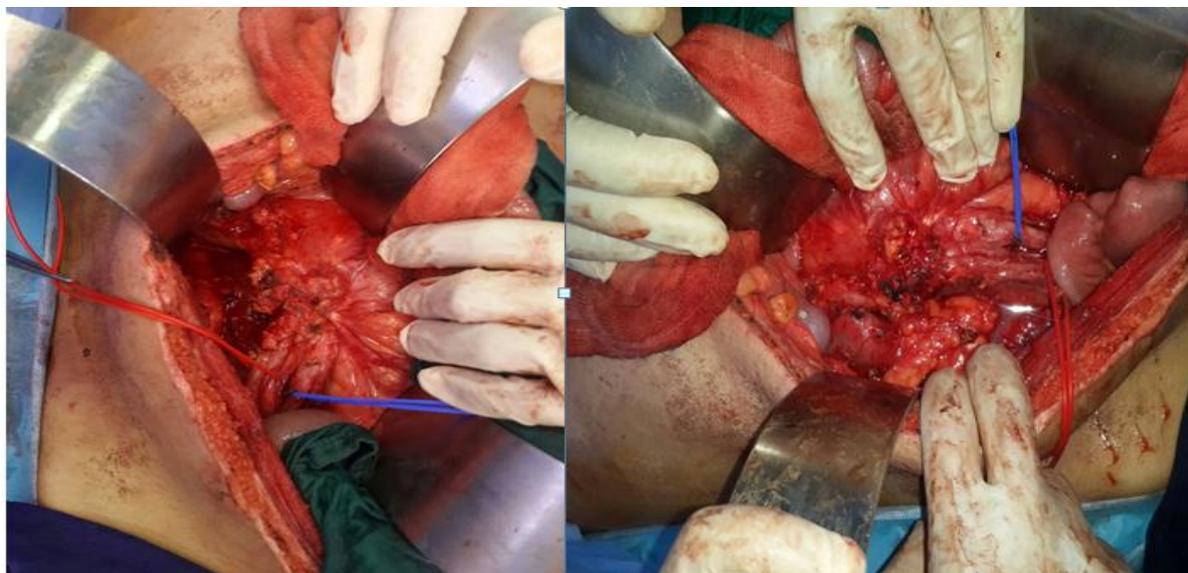


Figure 3: Picture showing failure of retroperitoneal lymphadenectomy.

Our patient died after 4 months of operation.

DISCUSSION

Paratesticular rhabdomyosarcoma accounts for 7% to 10% of all genitourinary tract RMS tumors and is the third most common following that of the prostate and bladder. The age distribution is bimodal, with peaks at 1 to 5 years and then at 16 years of age.^[1]

The consensus is that this tumor derives from mesenchymal elements of the testis envelope, epididymis and spermatic cord. The tumor manifests as a hard painless inguinoscrotal swelling, the size and duration of development are varied and it rarely invades the scrotal skin. The mass might evolve within the external inguinal ring away from the scrotal contents.^[2,3] A scrotal mass (85%) was the most common clinical presentation

followed by trauma or bruising (8%) and hydrocele or hernia (6%).^[1,9] On physical examination, a mass is usually palpable, however; in 15% to 20% cases the tumor can be associated with a hydrocele, which limits the testicular examination. Differential diagnoses include testicular torsion, orchiepididymitis, scrotal abscess and, rarely, testicular tuberculosis.

A testicular ultrasound is routinely performed for a scrotal mass. This imaging modality shows a mass with heterogeneous echogenicity and inguinoscrotal extension in 80% of cases.^[4] This allows the nature of the intrascrotal tissue mass to be determined and specifies the exact topography. Ultrasound is used to establish the differential diagnosis and eliminate diagnoses of simple cyst and varicocele.^[11,12]

A thoraco-abdomino-pelvic CT scan allows for any deep invasion of the lymph nodes to be investigated, especially lombo-aortic and pelvic metastases as well as possible metastases to the liver or lung. Most authors prefer the use of thoraco-abdomino-pelvic CT compared to ultrasound for lymphomas.

Magnetic resonance imaging is an efficient imaging modality when using surface coils. The tumor appears homogeneous in T1-weighted images and heterogeneous in T2-weighted images with signal intensity similar to that in a normal testis. The low signal intensity of the tunica albuginea in T2-weighted images allows the visualization of a clear separation of the mass from the testis.^[1,5,6]

In rhabdomyosarcoma, tumoral markers including alpha-fetoprotein, beta-human chorionic gonadotropin and carcinoembryonic antigen are usually normal. This was the case with our patient.

A malignant tumor might be suspected in masses sitting in the distal cord with a hard and irregular form adhering to surrounding structures; a rapid increase in tumor volume might be noted. But the diagnosis is mainly made using histology. The macroscopic features include a lobed tumor surrounded by whitish pseudocapsules, hemorrhagic array are sometimes revealed.^[7]

RMS has been traditionally classified into 3 main histologic subtypes: embryonal (80%), alveolar (20%), and pleomorphic.^[1,2,9] Additional variants have more recently been identified, such as spindle cell RMS and sclerosing RMS. Spindle cell and sclerosing RMS are rare, comprising only 5% to 10% of RMS.^[6,8,9]

Rhabdomyosarcoma may be included in the differential diagnosis for other paratesticular sarcomas such as leiomyosarcoma, liposarcoma and fibrosarcoma. However, these pathologies occur more often in adults. Imaging cannot discriminate between these tumors. The final diagnosis can be established by histological study after surgical excision of the tumoral mass.^[6,7]

Radical orchidectomy by the inguinal route with first cord ligation remains the essential act for histological diagnosis and constitutes the first step of treatment regardless of the stage of the disease. Hemiscrotectomy associating inguinal treatment is indicated first in scrotal cases whenever local invasion or presence of lymph are clinically evidenced.^[6,7] An inguinal lymphadenectomy should not be performed without first obtaining imaging, including a CT scan or lymphography.

Chemotherapy should be routinely administered since rhabdomyosarcoma is chemosensitive. This therapeutic approach consists of administering actinomycin D, vincristine and cyclophosphamide.^[5,6,7,10] Radiotherapy is a complementary treatment of chemotherapy and surgery to eliminate residual foci and retroperitoneal lymph nodes. Our patient benefited from an inguinal orchidectomy, chemotherapy and retroperitoneal lymphadenectomy. He died 4 months after.

CONCLUSION

Paratesticular rhabdomyosarcoma is a rare aggressive tumor manifesting in children and very young adults. Localized forms have a good prognosis whereas metastatic tumors show very poor results. A well-defined treatment based on surgery and chemotherapy yields good results. Radiotherapy is indicated in cases of residual foci and retroperitoneal lymph nodes. Strict follow-up has to be instituted for all patients.

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