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CLINICAL CASE

Primary testicular rhabdomyosarcoma: A case report☆

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KEYWORDS
Primary rhabdomyosarcoma;
Testicular sarcoma;
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Abstract
Background: Rhabdomyosarcoma is the most common sarcoma of soft tissues in childhood and adolescence, with an annual incidence of 4–7 cases per million children aged 15. Embryonal rhabdomyosarcoma is common in adults younger than 30 years, and are usually presented as a large painless, palpable mass (>5 cm). Survival in the case of paratesticular sarcoma in men is approximately 50%.
Clinical case: Male 27 years of age with no history of importance, was seen in a clinic with an increased, painless, left testicular volume 3 years onset. Intrascrotal left testicle increased volume, with dimensions of 20 cm × 12 cm × 8 cm, a stone and left inguinal node in induratum measuring 2 cm × 2 cm. Microscopically, it showed a pattern of an embryonal rhabdomyosarcoma with left inguinal node metastases.
Conclusion: Early diagnosis of testicular tumours, and especially of primary intratesticular rhabdomyosarcomas, and aggressive surgical treatment in combination with chemotherapy reduces the incidence of local recurrence and may improve the rate of disease-free survival and overall survival in adult patients with metastases.

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PALABRAS CLAVE
Rabdomiosarcoma primario;
Sarcoma testicular;
Tratamiento de rabdomiosarcoma

Rabdomiosarcoma testicular primario: reporte de un caso

Resumen
Antecedentes: El rabdomiosarcoma es el sarcoma de tejidos blandos más común en la infancia y adolescencia, con incidencia anual de 4 a 7 casos por millón de niños de 15 años de edad. El rabdomiosarcoma embrionario es común en adultos menores de 30 años, y se presenta usualmente como una masa indolora, palpable grande (>5 cm). La sobrevida en el caso de hombres con sarcoma paratesticular es aproximadamente del 50%.
Caso clínico: Paciente masculino de 27 años de edad, sin antecedentes de importancia. Acude a consulta por presentar 3 años con aumento de volumen testicular izquierdo, indoloro. A la exploración física del testículo izquierdo, es intraescrotal con aumentado de volumen, con dimensiones de 20 × 12 × 8 cm, pétreo y con ganglio en región inguinal izquierda indurado de 2 × 2 cm. Microscópicamente, con patrón de rabdomiosarcoma embrionario y ganglio inguinal izquierdo con metástasis de rabdomiosarcoma embrionario.
Conclusión: El diagnóstico temprano de los tumores testiculares, y en especial de los rabdomiosarcomas intratesticulares primarios, así como el tratamiento quirúrgico agresivo en combinación con la quimioterapia disminuyen la incidencia de recurrencia local y podrían mejorar la tasa de supervivencia libre de enfermedad y la supervivencia global en pacientes adultos con metástasis.

Background

Rhabdomyosarcoma is the most common sarcoma of soft tissues in childhood and adolescence, with an annual incidence of 4–7 cases per million children aged 15.[1] The aetiology of pure primary testicular rhabdomyosarcomas is unclear.[2,3] They are extremely rare, with only 14 cases having been reported in the literature.[3–8]

Embryonal rhabdomyosarcoma is common in adults under 30 years of age and usually present as a large painless, palpable mass (>5 cm). Testicular ultrasound imaging shows a solid mass, although at times no distinction may be made between benign and malignant tumours.

The procedure of choice is an inguinal approach with a wide incision and high ligation of the spermatic cord and the testicle. Few publications exist to report on the results of testicular rhabdomyosarcoma treatment. Six cases of primary testicular rhabdomyosarcoma in adults were recently published in which aggressive surgical strategies and chemotherapy were used, resulting in a reduction in local recurrence and an improvement in the disease-free survival rate and overall survival rate in adult patients with metastases.[8] There is an approximate survival rate of 50% in paratesticular sarcoma.[2]

The aim of this study is to present a clinical case study of primary testicular rhabdomyosarcoma in an adult, its aggression and management.

Clinical case

Male 27 years of age patient who presented at a clinic with an increased, painless, left testicular volume of 3 years onset. He also presented with a 6 month history of left hemi-abdominal pain, accompanied by nausea, immediately vomiting after meals and a weight loss of 12 kg during the last 2 months. On physical examination the patient had a Karnofsky performance score of 50, the abdomen had increased in volume due to the tumour which had spread to the left hemi-abdomen, it was palpably painful and adherent to deep tissue. The intrascrotal right testicle measured 4 cm × 3 cm × 2 cm, was of normal consistency, with normal spermatic cord; the intrascrotal left testicle had increased in volume, measured 20 cm × 12 cm × 8 cm, contained a stone and uneven edges. It moved within the testicular sac, was not palpably painful and the spermatic cord was normal to the touch. A left inguinal node in induratum measuring 2 cm × 2 cm was observed.

The study protocol for testicular tumours has pre-surgical alpha-fetoprotein (AFP) tumour markers of 1.39 UI/ml, human chorionic gonadotrophin beta (GCH-b) of 3.7 mUI/ml and lactate dehydrogenase of (DHL) 524 UI/l. A chest X-ray revealed no images suggestive of lung metastases or metastases in the mediastinum. Testicular ultrasound imaging is shown in Fig. 1. Simple CAT scanning of the abdomen and pelvis showed a paraaeortic conglomerated node which spread from the inferior mesenteric artery to the pelvic cavity, which led to severe pyelocaliceal ectasia of the left kidney (Fig. 2).

On 4th December 2014 a double j left stent was inserted and left radical orchiectomy was performed. During surgery we observed a left testicular tumour of 18 cm × 11 cm × 8 cm, of grainy consistency, with no invasion of the scrotum, with congested spermatic cord, and inguinal node induratum of 2 cm × 3 cm.

The official histopathological report stated that there was macroscopic evidence of a surgical specimen measuring 17.5 cm × 11 cm, coffee-coloured, with a neoplasic type whitish-grey 13 cm tumour inside it, h necrotic areas, and a 9 cm congested spermatic cord. Left inguinal node was 5.5 cm, grey in colour, with a neoplasic appearance. Microscopically, it showed a pattern of an embryonal