CASE REPORT

Rhabdomyosarcoma of spermatic cord in a 65-year-old man presenting as a groin swelling

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SUMMARY

A 65-year-old man presented with a swelling in the right groin of 6 months duration. The swelling was associated with dull aching pain and the patient reported of increase in size of the swelling during lifting of heavy weights. The swelling was 6×5 cm, hard in consistency, mobile and there was no impulse of cough. Ultrasonography showed a solid mass measuring 5.3×1.5×5.2 cm arising from the spermatic cord. High-inguinal orchiectomy was performed. Histopathology revealed rhabdomyosarcoma (RMS) of the spermatic cord. Patient was advised adjuvant chemotherapy but he refused. Spermatic cord RMS is a rare tumour derived from the undifferentiated mesoderm. It is most often observed in children and adolescents. It rarely appears after the second decade of life. It usually manifests as a painless, firm to hard mass in the inguinal canal or scrotum. Radical high-inguinal orchiectomy is the treatment of choice.

BACKGROUND

Paratesticular rhabdomyosarcoma (RMS) is a rare tumour. Though it is a tumour of the paediatric and adolescent age group, it should be considered in the differential diagnosis of groin swellings even in the elderly. It has a good prognosis in comparison with other sites of RMS. Hence surgery with adjuvant therapy has to be offered for disease free survival.

CASE PRESENTATION

A 65-year-old man presented with a swelling in the right groin of 6 months duration. The swelling was associated with dull aching pain. The patient reported increase in size of the swelling during lifting of heavy weights. The groin swelling was 6×5 cm, hard in consistency, mobile and there was no impulse of cough (figure 1). There were no

other swellings in the body. The patient was provisionally diagnosed to have a solitary neurofibroma of right inguinal canal. Ultrasonography was performed. The patient was taken up for inguinal exploration. On exploration, a globular, grey white, capsulated soft tissue mass (measuring 5×2×5 cm) with bosselated external surface was seen (figure 2). A high-inguinal orchiectomy was performed (figure 3). Histopathology revealed RMS of the spermatic cord. After consultation with multidisciplinary team adjuvant chemotherapy with vincristine, dactinomycin, and cyclophosphamide (VAC) was advised. The patient was explained about the number of cycles of chemotherapy, its complications and the risks involved. The patient declined the chemotherapy citing his old age.

INVESTIGATIONS

Ultrasonography showed a solid mass measuring 5.3×1.5×5.2 cm arising from the spermatic cord.

DIFFERENTIAL DIAGNOSIS

- ▶ Lipoma
- ▶ Neurofibroma
- ▶ Desmoid tumour
- ▶ Primary lymphoma
- ▶ Sarcoma
- ▶ Metastases to the inguinal canal

TREATMENT

High inguinal orchiectomy

Adjuvant chemotherapy with VAC was advised, but patient refused.

OUTCOME AND FOLLOW-UP

The patient is currently disease free for the last 3 years and is on regular annual follow-up.

Figure 1 Swelling in the right inguinal region measuring 6×5 cm.

Figure 2 Rhabdomyosarcoma arising from the spermatic cord on inguinal exploration.

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Figure 3 Gross specimen showing globular, grey white, capsulated soft tissue mass (measuring 5×2×5 cm) with bosselated external surface. On cut section—grey white, lobulated with foci of haemorrhagic areas.

DISCUSSION

RMS is defined as a malignant mesenchymal tumour of the muscle tissue, commonly occurring in the head and neck, extremities and genitourinary tract.¹ Paratesticular RMS, which includes RMS of the spermatic cord, epididymis, testis and testicular tunica accounts for 7% of all RMS.² Incidence of these tumours is highest in the younger age groups, with two frequency peaks—one at around 4 years of age, and another, at around 18 years of age. In fact, RMS is the most common soft tissue sarcoma of childhood.³ It rarely appears after second decade.⁴

Clinical features of this tumour depend on the site of tumour. Paratesticular RMS generally tends to be aggressive, and may present as hard painless unilateral swelling of the inguinoscrotal region. However pain may be present in some cases, leading it to be often mistaken for epididymitis.² Metastasis is not uncommon, and typically occurs by haematogenous or lymphatic route. Most common sites of metastasis include regional nodes, lungs and cortical bone.⁵ Imaging that is most commonly used in the diagnosis of paratesticular RMS is scrotal ultrasonography, which is non-specific, showing a mass with heterogeneous echogenicity and inguinoscrotal extension in the majority of cases.²

Diagnosis of the condition is histopathological. Grossly, the tumour is lobed, and pseudoencapsulated. Microscopically, the tumour can be identified by the presence of rhabdomyoblasts. Stains used to identify the tumour include desmin stains, muscle-specific actin and myogenin. There are five recognised variants of RMS as per international classification: Embryonal, botryoid embryonal, spindle cell embryonal, alveolar and anaplastic. Alveolar is the most common of these variants and is often seen in the head and neck as well as the genitourinary tract.⁶

One point of view held almost unanimously by authors is that early detection of this condition is critical. The most effective primary treatment modality in such a case is radical orchiectomy. If there is fixation of tumour to the scrotal wall, hemiscrotoectomy is an additional procedure to be performed. In case of suspected nodal involvement, CT of pelvis can be used to confirm, before inguinal lymphadenectomy.^{2–3} However, local recurrence is frequent. Hence follow-up with adjuvant chemotherapy and radiotherapy is a must.⁷

The International Rhabdomyosarcoma Study Group has classified postsurgery RMS as per the text below⁸:

Group 1: localised disease, completely excised, no microscopic residual

A. Confined to site of origin, completely resected

B. Infiltrating beyond site of origin, completely resected

Group 2: total gross resection

A. Gross resection with evidence of microscopic local residual

B. Regional disease with involved lymph nodes, completely resected with no microscopic residual

C. Microscopic local and/or nodal residual

Group 3: incomplete resection or biopsy with gross residual

Group 4: distant metastases

Chemotherapy is very useful. It is often used in tandem with radiotherapy. Most common chemotherapeutic drug combination is VAC regimen. In some cases, chemotherapy may be sufficient on its own in clearing residual metastatic lesions postsurgery. Ifosfamide—etoposide combination has also been shown to be just as effective.⁸ Radiotherapy is indicated mainly for residual foci and cases with retroperitoneal lymph nodes.⁹

Thus, the gold standard of treatment of non-metastatic embryonal RMS is combination of surgery with adjuvant chemotherapy VAC and radiotherapy.⁸

Primary paratesticular RMS is considered to have a good prognosis in comparison with other sites of RMS.⁹

Learning points

- ▶ Paratesticular rhabdomyosarcoma (RMS) is commonly seen in children and adolescents, and rare in adults.
- ▶ The primary paratesticular site is considered to have a good prognosis in comparison with other RMS.
- ▶ Radical high-inguinal orchiectomy followed by adjuvant therapy is the treatment of choice.

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Competing interests None.

Patient consent Obtained.

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