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A Case Report

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Recurrent Para -Testicular Rhabdomyosarcoma: A Case Report

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Abstract

Background: Para-testicular RMS is not frequently reported and originates from the stromal tissues of the spermatic cord, epididymis, testis and tunica. It is a very uncommon feature in RMS and may be seen in a small percentage of patients and most often in children. At times the diagnosis may pose as a problem and physical examination may be inadequate at which time ultrasonography comes in handy. The main form of management is surgical through radical orchiectomy and the medical form is through Actinomycin D, vincristine and cyclophosphamide and very rarely radiotherapy.

Keywords: Para-testicular rhabdomyosarcoma,Radical orchiectomy,Chemotherapy,Ultrasonography

Introduction:

There are following sites of origin for Para-testicular RMS: spermatic cord; epididymis; testis and tunica vaginalis. It is noted in 7 % of all patients in IRS and accounts for 17% of all malignant intrascrotal tumours in children less than fifteen years of age [1]. Alveolar; Embryonal; Botryoid embryonal; Spindle cell embryonal; Anaplastic These are according to the internation RMS classification. rRMS is the most frequent variant, encompassing 60% of cases; it is also known as embryonal RMS (eRMS). Para-testicular rhabdomyosarconi is often associated with rather vague clinical symptoms. The diagnostic procedures are – ultrasonography (US) which is used to identify the tumor localization if it cannot be determined by physical examination [3]. For histological diagnosis and the initial treatment, the initial surgical procedure is radical orchiectomy through the inguinal route ligated initially irrespective of the disease state. Like most adult soft tissue sarcomas, rhabdomyosarcoma is chemosensitive; this is why chemotherapy forms a section of the treatment regimen; typical drugs are Actinomycin D, vincristine and cyclophosphamide. Radiotherapy is also sometimes incorporated in this treatment [3].

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Case Presentation:

In April 2023 a 16-year-old boy presented in Surgical OPD with swelling in the right inguinal region of one months duration, who had right radical orchiectomy done one year back for scrotal mass biopsy revealed embryonal rhabdomyosarcoma of para testicular origin while testes and speramtic cord were negative for tumor. He had completed adjuvant chemo-therapy of six cycles of VCR 2 gm, CTX 1 gm and DOX 90 mg. There we saw the scar at the operation site in the right inguinal area and a hard irregular rubbery immobile mass which extended up to the scrotum. Ophthaloscopic examination was also unproductive; transillumination test was also negative. Routine investigations were normal and so was the liver function whereas, tumor markers including LDH, alpha-fetoprotein and beta-HCG were well within normal range. On the abdominal ultrasound there was a lobulated mass of size 5x9 cm in the right inguinal region that had micro-calcification. On the abdomen and pelvic CT scan, a deforming mass of size 10cm X 6 cm was observed in the right inguinal region with fat stranding and enlarged nodes were also noted and multiple indeterminate pulmonary nodules were also observed (Fig 1 & 2). A tru-cut biopsy was performed and immune staining with desmin and myo-D was positive as shown in figures 3 and 4 and the diagnosis of recurrent embryonal rhabdomyosarcoma was made and confirmation was sought from other pathologists. A preoperative patient was on the stage. In course of surgery, a massive necrotic area was seen involving the adductor muscles and the lymph nodes above and below the oblique abdominal muscle; the scrotum was also found to have necrosis. The mass was felt to be misconceived as non-resectable and consequently debulking was done (Figure 5). The specimen was finally subjected to histopathology as the one depicted in the figure 6. He was subsequently referred to a medical oncologist for assessment and management of the case.

Figure 1: Ct Abdomen Pelvis Shows A Large Right Inguinal Region Mass As Pointed By The Red Arrow

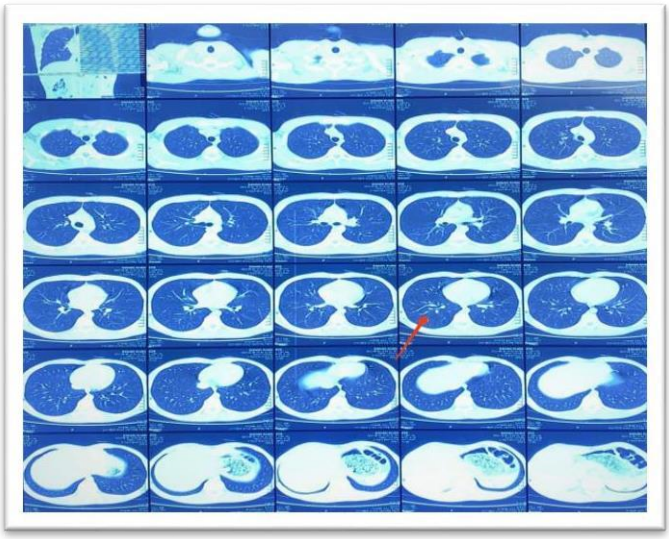
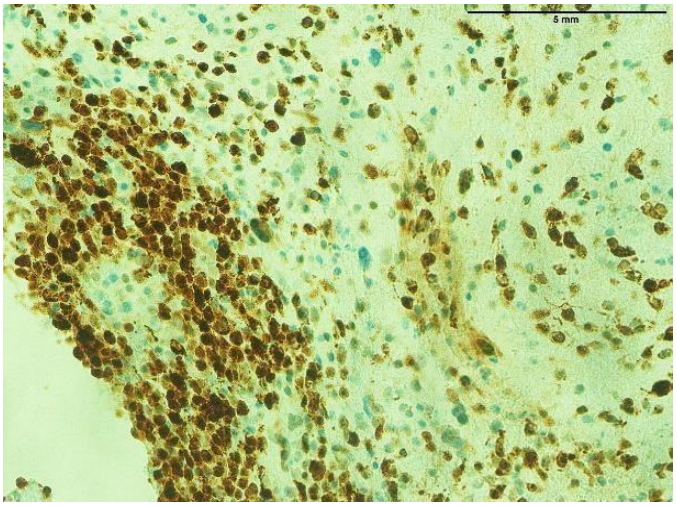


Figure 2: Ct Chest Shows Some Indeterminate Pulmonary Infiltrates As Pointed By The Red Arrows



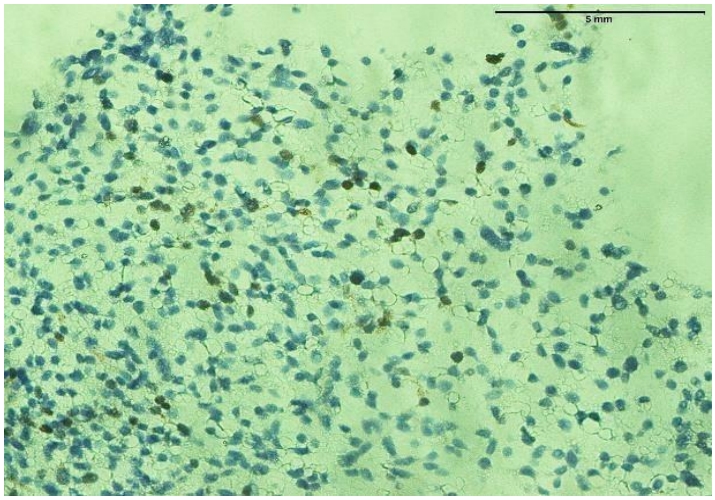


Figure 3: Desmin Positive
Figure 4: Myogenin D Positive

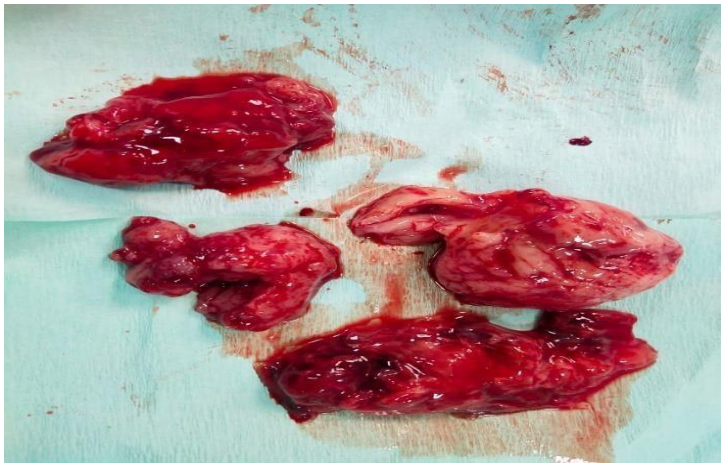


Figure 5: Debulked Tissues Of The Tumour

Discussion

Alveolar rhabdomyosarcoma (RMS) serves from embryonal mesencies and is one of the most malignant tumors. It has four subtypes: These areembryonal, alveolar, pleomorphic and spindle cell. RMS typically arises from soft tissues and affects mainly the head and neck, ocular and orbital region, GU tract and genitals, and limbs. Testicular RMS is also known as para – testicular RMS and is the seventh most prevalent RMS responsible for 7% of all RMS; it is a rather rare tumor that develops from spermatocytic cells of epididymal and spermatic cord origin. This tumor is seen in children and young adults and therefore the age distribution is bimodal with age of 5 and 16 respectively as in the above case. Depending on the stage of development the RMS can be classified into embryonal RMS and it is the most common; the disease can be cured if identified in its early stage. Nevertheless, the relapse rate is quite high, and in our patient, recurrent bilateral pulmonary nodules were diagnosed one year after right orchiectomy. Most para-testicular tumours are found incidentally and are painless mass in the scrotum; at the time of presentation, they have often spread hematogenously to iliac and paraaortic lymph nodes or lungs and bones [3]. The oedema is asymptomatic and so patients do not seek medical help and end up presenting at the advanced stage of the disease. The manoeuvres that should be included are the scrotal palpation, inguinal lymph node, other lymph node, general physical examination and the systemic examination for metastasin. Para_testicular RMS is amongst the causes of scrotal swellings and treatment with inguino_scrotal ultrasound is useful in differentiating it from other conditionsdue to the demonstration of the lobulated heterogeneous mass that characterizes this tumor vulgaris. Tumor marker assays that are normally elevated in testicular swellings are usually normal in para-testicular RMS although there may be a slight rise in the LDH [6]. For staging of disease and to check loco-regional invasion or metastasis, Thoraco-Abdomino-Pelvic CT and MRI are the gold standard. All these imaging techniques assists in planing the treatment [7]. Treatment of para-testicular RMS should consist of surgery – radical, retroperitoneal lymph node dissection, radiotherapy and chemotherapy [8]. In radical orchiectomy essential histological information is obtained and this is the first operation carried out. This is then followed by chemotherapy, which normally takes 4-6 cycles of a program such as Vincristine, Cyclophosphamide and Adriamycin (VAC), which is known to be effective [9]. Regional

lymph node and metastatic disease is mainly treated by radiotherapy. For the children affected by the para-testicular RMS the prognosis is comparatively better than that of the adults, therefore, early diagnosis and multifaceted treatment is advisable [10]. Such patients therefore require follow up and regular evaluation, in order to note the re occurrences and any consequent effects. It becomes important to rise the level of attention and to detect the disease in its early stages to improve the prognosis and survival rate of the para-testicular R-Med patients.

Conclusion

As mentioned earlier, we suspect that the diagnosis of para testicular RMS is not straightforward in many cases and ultrasonography becomes particularly beneficial if physical examination only is inconclusive. The management consists of the surgical treatment that is the removal of the tumor with the affected testicle, orchiectomy; the chemotherapeutic treatment usually Actinomycin D, vincristine, cyclophosphamide and the use of radiotherapy in some special cases.

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