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CASE REPORT

Alveolar Rhabdomyosarcoma Of Paratesticular Tissue In An Adult

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ABSTRACT

Most masses encountered within the scrotal sac are in the testis and are neoplastic. However, asubset of these tumours are extratesticular and arise from the paratesticular tissue. The paratesticular region is a complex anatomical area which includes the contents of the spermatic cord, testicular tunics, the epididymis and the testis [1]. Histogenetically, this area is composed of a variety of epithelial, mesothelial and mesenchymal elements. Rhabdomyosarcoma of the male genital tract is rare. If present, it is usually seen in children and adolescents and is of the embryonal type. We present here, a case of alveolar rhabdomyosarcoma in an elderly patient.

Key Words: Rhabdomyosarcoma, Paratesticular tissue, Adult

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Introduction

Rhabdomyosarcoma is the most common soft tissue sarcoma of childhood, representing 5% of all childhood cancers [2]. It is thought to arise from primitive mesenchymal cells which are committed to skeletal muscle differentiation and can occur in a variety of organs and tissues, including those that lack striated muscle. Rhabdomyosarcoma represents 24% of all adult paratesticular tumours. 80% of rhabdomyosarcomas occur before the age of 21yrs, with the remaining 20% evenly spread throughout the remaining decades [3],[4]. Histologically, any subtype of rhabdomyosarcoma may occur in the paratesticular region. Embryonal rhabdomyosarcoma is the commonest variety of tumour in the paratesticular region. Cytogenectically, the partial monosomy of chromosome 11 often characterizes embryonal rhabdomyosarcoma. A variant of embryonal rhabdomyosarcoma i.e. spindle cell rhabdomyosarcoma deserves special mention because it has an excellent prognosis. Alveolar rhabdomyosarcomas are less common than the embryonal subtypes, mainly occur in adolescents and have a bad prognosis. Two consistent translocations have been found in the majority of patients with alveolar rhabdomyosarcoma, on applying PCR FISH on their samples. The first, t(2;13)(q35;q14), results from the fusion of the PAX3 and FKHR genes, while the second, t(1;13)(p36;q14), involves the fusion of FXHR and PAX7. In embryonal rhabdomyosarcoma, loss of heterozygosity at 11p15.5 has been identified. In addition, the alveolar subtype is diploid or hypodiploid, whereas the embryonal variety usually demonstrates tetraploidy [5],[6].

Pleomorphic rhabdomyosarcoma is the rarest type, occurs mainly in adults and has a poor prognosis.

A case of alveolar rhabdomyosarcoma is documented here, due to its rarity and the effect on its treatment and prognosis.

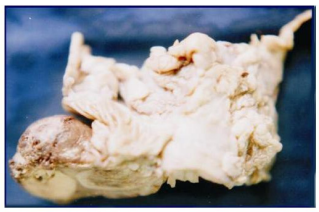
Case Presentation

A male aged 55yrs presented with complaints of a painful, gradually increasing right scrotal mass for 6 months. On clinical examination, a nodular, firm mass was felt in the caudal region of the epididymis. USG revealed a hetrogenous round tumour formation. All preoperative analysis including his x-ray and abdominal ultrasound were normal.

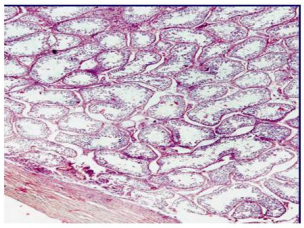
Inguinal orchidectomy, with high ligation of the spermatic cord (radical orchidectomty) was performed and the specimen was sent to the department of pathology for HPE.

GROSS-the right side testis with sac was received. On opening the sac, serous fluid came out. The wall was thickened. The cut surface was homogenous brown.

The spermatic cord showed three nodules measuring 2x2cm, 1x0.5cm and 0.5x0.5cm in size, which were well encapasulated. The cut surface of each nodule was homogenous brown [Table/Fig 1].



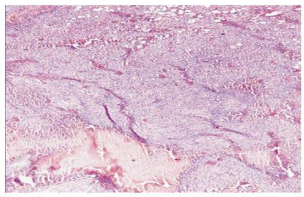
(Table/Fig 1)Specimen showing thickened sac, nodular surface with areas of normal testis.



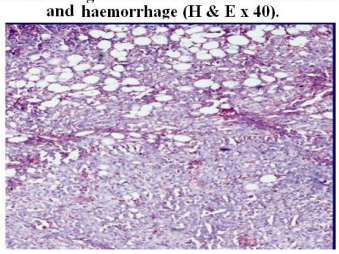
(Table/Fig 2) Section showing normal tubules with spermatogenesis (H& E x 40)

Microscopic

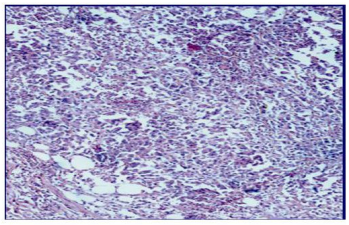
The section from the testis showed normal structure with complete spermatogenesis upto the spermatozoa stage [Table/Fig 2]. The sections from the nodules in the cord showed collections of cells in the alveolar and solid patterns. The cells were pleomorphic and hyperchromatic, with moderate eosinophilic cytoplasm, eccentric vesicular nucleus and prominent nucleoli. Numerous mitotic figures, multinucleated cells and areas of necrosis were also present. In between, interspersed fibrous tissue infiltrated by inflammatory cells were also seen [Table/Fig 3],[Table/Fig 4],[Table/Fig 5],[Table/Fig 6],[Table/Fig 7].Vascular invasion was not present. A provisional diagnosis of alveolar rhabdomyosarcoma on the basis of morphology and immunohistochemistry for Vimentin, Desmin, Myogenin and MyoD, was suggested.



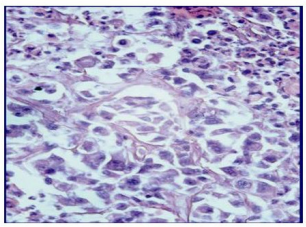
(Table/Fig 3) Section showing diffuse tumour cells infiltration with necrosis and haemorrhage (H & E x 40).



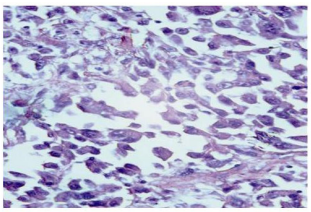
(Table/Fig 4) High power view of (above image) (H & E x 100).



(Table/Fig 5) Section showing tumour cells in alveolar pattern, solid pattern, (H & E x 100).

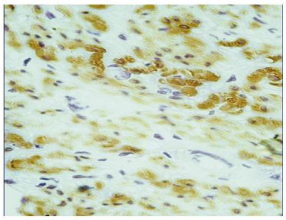


(Table/Fig 6)Section showing pleomorphic tumour cells, in alveolar pattern, mitotic figures, (H & E x 400).



(Table/Fig 7) Section showing tumour cells, mitotic figures, multinucleated giant cells (H & E x 400).

Our case showed positivity for Desmin [Table/Fig 8]. The patient could not get the other markers done as he was very poor.



(Table/Fig 8) IHC of the biopsy showing desmin positivity (IHC x 100).

Discussion

The other tumours which can also present as paratesticular tumours, are Malignant mesothelioma, Malignant lymphoma and Metastatic tumour.

The usual age for the incidence of malignant mesothelioma is in between 55–75 years. Clinically, the patients usually present as a firm,painless scrotal mass in association with a hydrocoele. Grossly, there is the thickening of the tunica vaginalis with multiple friable nodules or excrescences. Histologically, about 75% cases are purely epithelial. They have either papillary or tubulopapillary morphology. The cells are arranged in solid nests, have prominent nucleoli and the cytoplasm is eosinophilic. Immunohistochemically, they are positive for AE1/AE3, Calretrin, EMA and Vimentin. Malignant mesothelioma was ruled out on the basis of morphology and immunohistochemistry.

The usual age for the incidence of malignant lymphoma is in between 21-89 years. These tumours clinically present as painless hard masses. Histologically, there is a monomorphic population of cells in diffuse sheets. These cells had hyperchromatic, pleomorphic nuclei, prominent nucleoli and a scant amount of cytoplasm. Immunohistochemically, they are negative for Vimentin, Myogenin and MyoD.

The paratesticular region may occasionally be the site of metastatic lesions. The commonest site of the origin of primary tumours metastasising to the paratesticular region are stomach, prostate and kidneys, which were ruled out by the lack of specific morphology[7,8].

Conclusion

This case emphasizes the importance of consideration of alveolar rhabdomyosarcoma in diagnosing tumours of paratesticular tissue. The case also emphasizes to consider this diagnosis in an adult.

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