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Embryonal Rhabdomyosarcomas of the Testis, Our Experience in Medical College Hospital

KEYWORDS: Rhabdomyosarcomas, Paratesticular tumors, Retroperitoneal lymphadenopathy, Orchidectomy.

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ABSTRACT *Rhabdomyosarcoma is not only the most common soft tissue sarcoma in children under fifteen years of age, but also one of the most common soft tissue sarcomas of adolescents and young adults.1*

It is rare in persons older than 45 years. We report two cases of rhabdomyosarcomas presented to the department of surgery, Kurnool medical college, Kurnool.

Introduction:
Three percent of childhood cancers are soft tissue sarcomas and one percent of adult cancers are soft tissue sarcomas. Rhabdomyosarcomas arising from mesenchymal cells, is the most common soft tissue tumor in children and accounts for fifty percent of sarcomas.[2] But, the incidence of rhabdomyosarcomas in adults is rare, accounting for only three percent of soft tissue sarcomas.[3] It is an intra scrotal tumor that is localized in paratesticular structures such as epididymis, spermatic cord, and mesenchymal elements of the testis. It represents seven percent of all adult rhabdomyosarcomas according to the Inter group rhabdomyosarcoma study.[4] Typically, rhabdomyosarcoma presents as a painless scrotal mass.

Case one:
A 18 year- old male patient presented to the surgical department of Kurnool medical college, Kurnool, with a three month history of painless, left scrotal swelling, increasing in size.

Clinical examination reveal hard testicular mass in his left side with a diameter of 12cm. An ultrasound reveals a large heterogeneous tumor mass in the left testis. CT scan reveals paraaortic lymphadenopathy.Tumor markers were within normal limits.

Patient underwent left radical orchidectomy and specimen was received in the pathology department. The size of the specimen was 11X6X6cm (fig1).

Case two:
A 13 year- old male patient presented to the surgical department of Kurnool medical college, Kurnool, with a two month history of painless, left scrotal swelling, increasing in size.

Clinical examination reveal hard testicular mass in his left side with a diameter of 11X6X5cm. An ultrasound reveals a large heterogeneous tumor mass with calcifications in the left testis. CT scan reveals paraaortic lymphadenopathy.Tumor markers were within normal limits.

Patient underwent left radical orchidectomy and specimen was received in the pathology department. The size of the specimen was 11X5X4cm Immunohistochemistry demonstrated expression of myogenic antigens,supporting a greater degree of differentiation of spindle-shaped cells(fig 2).Tumours of both patients were positive for Vimentin(fig 3).

Discussion:
Rhabdomyosarcoma,inspite of diversity in location,clinical presentation, and histologic picture,it has uniform age incidence;it occurs predominantly in infants and children and less frequently in adolescent and young adults.Horn and Enterline revised the first classification scheme based on the clinical and pathologic features of these tumours. They identified four subtypes. Embryonal,Botryoid,alveolar and pleomorphic.

Each of these Rhabdomyosarcomas subtypes occurs in a characteristic age group.For example embyonal Rhabdomyosarcoma and botryoid and spindle cell subtype affects mainly but not exclusively,childres between birth and 15 years of old age.On the other hand,alveolar rhabdomyosarcoma tends to affect older patients, with a peak age of 10-25 years.Rhabdomyosarcoma may arise anywhere in the body.Three regions are particularly affected: the head and neck most commonly affected,genitourinary tract and retroperitoneum and upper and lower extremities in that order.

In the head and neck parameningeal tumours are the most common and orbit is the next most common site.Histologically embryonal subtype is the most common.

Genitourinary tract is the second most common site for Rhabdomyosarcoma.71% of patients in the IRS I and IRS II with genitourinarary tumours were of embryonal subtype, and 21% were of botryoid subtype;and alveolar subtype of rhabdomyosarcoma was rare.[5]

The spindle cell subtype of Rhabdomyosarcoma has a propensity to arise in the paratesticular area.[6]Tumours of par-

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atesticular region are usually unilateral,painless mass presents for few weeks and months and located at upper pole of the testis.They are usually separate from the testis proper.These tumours have increased incidence of involvement of para aortic and retroperitoneal lymphnode involvement.[7]

Histologically, embryonal rabdomyosarcoma resembles various stages of embryogenesis of normal skeletal muscle. It is characterised by varying degrees of cellularity with alternating hyper cellular areas and myxoid areas. It composed of atypical spindle – shaped cells with scattered elongated rhabdomyoblasts. Spindle cell subtype is characterised by relatively uniform spindle – shaped cells deposited in a myxoid stroma. Cells are arranged in an irregular fascicular pattern reminiscent of leiomyosarcoma. Botryoid type of rhabdomyosarcoma is characterised by myxoid appearance and the submucosal hypercellular "cambium" layer. Alveolar rabdomyosarcoma is characterised by fibro vascular septa and alveolar growth pattern.

Conclusion:

Paratesticular rhabdomyosarcoma is a rare, aggressive tumor and grows rapidly in children and young adults. Prognosis depends on the stage of the tumor, age at diagnosis is an important, independent predictor of outcome. A localised form carries good prognosis, whereas metastatic tumor carries bad prognosis. Prognosis also depends on histological type.An alveolar subtype carries poor prognosis than the embryonal subtype. Early recognision of the tumor is the key for the best outcome. Surgery, sugery plus chemotherapy provides good results. When retro peritoneal lymphnodes, para aortic lymphnodes are involved, radiotherapy is given as an adjuvant therapy.

FIGURE 1

c/s grey white solid growth 9x5x4cms with myxoid glistening surface, focal hemorrhagic area & peripherally placed testis of size 3x2cm.

Figure 2

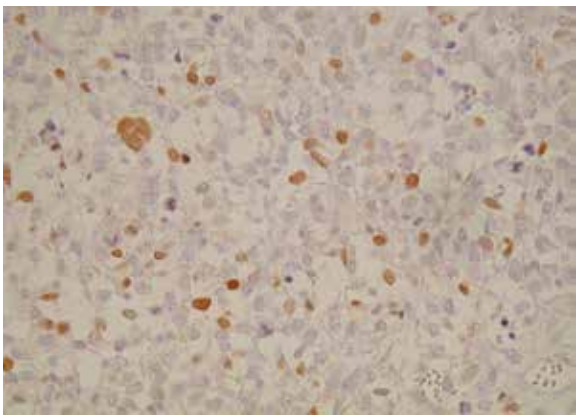

Figure 3 vimentin positive

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