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Rare case of paratesticular rhabdomyosarcoma

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Abstract Extremities are the usual site of involvement in Rhabdomyosarcomas whereas paratesticular rhabdomyosarcoma is an extremely rare, which is known to be an aggressive variety of paratesticular tumors occurring in first and second decades of life. They usually develop from the tunica, epididymis or the spermatic cord. Precise preoperative diagnosis is seldom established. It can be mistaken as epididymitis, scrotal abscess, hydrocele, testicular tumors, tuberculosis of testis and strangulated or obstructed inguinal hernia, as in our case. Multidisciplinary approach is needed for the management and disease localized to the scrotum has a favorable outcome.

Keywords Paratesticular tumors; rhabdomyosarcoma; child; orchidectomy.

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INTRODUCTION

Paratesticular rhabdomyosarcoma is an extremely rare tumor, and aggressive variety of paratesticular tumors having two peaks of incidence, most commonly occurring between 1st to 5th years of life [1]. It is very rare that a correct preoperative diagnosis is made as the condition is not common and

usually confused with epididymitis, scrotal abscess, hydrocele, testicular tumors, tuberculosis of testis and strangulated or obstructed inguinal hernia. Ultrasonography may be helpful preoperatively to differentiate intratesticular lesions from extratesticular lesions. Multimodality treatment and limited disease have better outcomes. Combined approach with surgery, chemotherapy and radiotherapy enhance the cure rate. We present a rare case of one year old boy with left paratesticular rhabdomyosarcoma misdiagnosed as obstructed inguinal hernia and without orchidectomy due to refusal of consent doing well on regular follow up without any recurrence after two years.

CASE REPORT

One year old boy presented to the emergency department with mother complaining of swelling in the left side of scrotum since last 4 months with pain and tenderness since three day associated with two episodes of bilious vomiting and fever. There was history of absolute constipation since one day. On examining the patient pulse was 124 per minute and was febrile. Abdomen was distended, soft and absent bowel sounds. X- ray of the abdomen did not reveal any air fluid levels. On examination there was left scrotal swelling

of approximately 6 cm to 4 cm in dimensions, was with reddened overlying skin, which was tender to touch. There was no visible peristalsis or impulse on crying. Opposite side examination was unremarkable. A clinical diagnosis of obstructed inguinal hernia was made and patient was prepared for exploration.

On exploration there was no hernia but a 4 cm x 3.5 cm X 3 cm firm encapsulated mass, grey- white in colour, completely separate from the testis arising from the lower end of the spermatic cord was found. En bloc excision of the mass with preservation of the left testis was done. On cut section there were small hemorrhages at the centre of the mass and glistening grey-white to grey yellow areas (Fig. 1) the mass was sent for histological examination and patient discharged on day 4th and advised to follow up.



Fig. 1. Cut section of the paratesticular mass after excision.

Histological examination was suggestive of pleomorphic variety of paratesticular rhabdomyosarcoma. Mitotic figures were evident (5-6/10 hpf) with strap cells, spindle cells and tadpole cells along with some cells with bizarre nuclei. Immunohistochemical examination was positive for desmin. The parents of the child were counseled for completion orchidectomy for which they refused. Workup for metastasis was done, which was normal without any involvement of retroperitoneal lymph nodes and the patient was given adjuvant chemotherapy. (Vincristine, adriamycin and cyclophosphamide). Patient was regularly followed up thereafter and there is no recurrence until two years of follow up.

DISCUSSION

Rhabdomyosarcomas are one of the commonly occurring soft tissue sarcomas of childhood; however paratesticular rhabdomyosarcoma is rare and consists 7% of all rhabdomyosarcomas (20 of 289 children in Intergroup Rhabdomyosarcoma Study). They develop from the mesenchymal tissue of tunica, epididymis or spermatic cord [1]. There are two peaks of incidence, most commonly between 1st to 5th years of life, with first peak in infancy followed by a second in adolescence [1,2]. Without any specific predilection for race.

Various histological varieties of paratesticular rhabdomyosarcoma are pleomorphic, alveolar, botryoidal, mixed tumors and embryonal [1]. Pleomorphic is the rarest type with poor prognosis and seen usually in adults and embryonal variety is common seen in young and has the best prognosis [3]. Rhabdomyosarcoma arises from primitive mesenchyme of the fetus with characteristics of striated muscle, immunohistochemical expression is positive for myosin and actin, desmin, myoglobin, and Z-band protein [4].

Clinically these tumors present as painless inguinoscrotal swelling and can be mistaken for conditions like epididymitis, hydrocele, scrotal abscess, testicular tumors, tuberculosis of testis, strangulated or obstructed inguinal hernia, as in our case [5]. However painful swellings are not commonly seen and have been reported in only 7% of cases [3]. Paratesticular masses are usually diagnosed in adolescence as a hard mass superior to the testis. On the physical examination mass appears to be separate from the testis however it is not always the case and sometimes it becomes difficult to demonstrate it as a separate mass. Paratesticular rhabdomyosarcoma spreads to the lungs, lymph nodes and bones with a high likelihood for the involvement of retroperitoneal lymph nodes [6]. This

increased incidence of retroperitoneal lymph node spread in paratesticular rhabdomyosarcoma favors the rationale of radical orchidectomy followed by evaluation of the status of retroperitoneal lymph nodes. Children over ten years of age have increased risk of nodal spread and relapse [7].

To differentiate paratesticular tumor from conditions mimicking it one should have high index of suspicion. Scrotal ultrasonography is the most valuable imaging modality showing its origin extratesticular or intratesticular and echo poor mass with or without accompanying hydrocele. Age and gradual onset of testicular mass are the key points helpful in differentiating paratesticular tumor from epididymitis [8]. Contrast enhanced CT scan of abdomen and pelvis with thin cuts is valuable to assess the retroperitoneal lymph node involvement and staging the disease, which is reported to be in tune of 25% to 30% in the literature [9,10].

Localized tumors limited to the scrotum have been seen to carry good prognosis. Modern day approach for the management of paratesticular rhabdomyosarcoma is multidisciplinary including surgery, chemotherapy with or without radiation therapy. Surgery includes high inguinal orchidectomy with excision of tumor and

hemiscrotectomy if there is involvement of scrotal skin. Surgery followed by chemotherapy is a standard due to highly chemosensitive nature of these tumors. Radiotherapy is administered in the presence of pathological retroperitoneal lymph nodes as an adjuvant. Retroperitoneal lymphnode dissection is not routinely done as microscopic nodal disease is effectively dealt with chemotherapy and radiotherapy. This aggressive combined approach has greatly enhanced the overall survival rate and with reduction in complication rate [11]. There has been a drastic reduction of cure rates from 25% to 70% since inception of multimodality treatment [12]. Highly chemosensitive nature of the tumor has decreased the role of surgery and radiotherapy in early stages.

Conclusion

Paratesticular rhabdomyosarcomas are very rare aggressive tumors of childhood and adolescence. It can mimics as epididymitis, scrotal abscess, testicular tumors, tuberculosis of testis, hydrocele, obstructed or strangulated hernia so high index of suspicion has to be kept in mind. Multimodality treatment is very effective and limited disease has better outcomes whereas metastatic disease has very poor outcome. Children less than ten years of age have decreased risk of nodal spread and

relapse. There has been drastic reduction in the cure rates since the inception of multimodality therapy.

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malignant mesenchymal tumors studies

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