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Testicular tumours in prepubertal children: About eight cases

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ABSTRACT

Background: To analyze the spectrum of testicular tumors in prepubertal children and the therapeutic resultants in an unselected population. **Materials and Methods:** Our hospital database was analyzed for testicular tumors from January 1995 to December 2010 concerning clinical presentation, treatment and therapeutic results. **Results:** Eight patients were operated on because of testicular tumors. In six cases (75%) the tumor was benign: benign teratoma (four cases), epidermoid cyst (one case) and immature teratoma (one case). Two patients (25%) had a malignant tumour: yolk-sac tumour (two cases). All this children underwent surgery. Radical inguinal orchidectomy was performed in six cases and conservative surgery was performed in two cases. One patient has received adjuvant chemotherapy. Follow-up was uneventfully three years after primary surgery. **Conclusion:** In prepubertal children, most testicular tumours are benign. If tumour markers were negative testis-preserving surgery can be proposed, complete excision of the tumour should be ascertained. In the case of testicular teratoma, the possibility of contralateral tumour should be considered in the follow-up.

Key words: Children, Testis, Tumour, Teratoma, yolk-sac tumour

INTRODUCTION

Testis tumours are rare in children compared with those occurring post-pubertally or with other genitourinary tumours in childhood. The annual incidence of testis tumour in children is only 0.5-2 per 100,000 boys, accounting for 1-2% of all paediatric tumours. They are divided into two main groups: germ cell tumour (GCT) and nongerm cell tumour (NGCT). There is a bimodal age distribution of the incidence of testicular tumours, in fact, they can occur in all age groups with two peaks: before three years and at the postpubertal period.[1,2]

Benign lesions represent a greater percentage of cases in children than in adolescent patients or in adults. Twenty of the 26 (77%) patients younger than 13 years reported by Taskinen were benign. In contrast, only 38% of postpubertal children have benign tumour, and all cases of yolk-sac tumours occurs in preschool-aged children.[3,4]

There is still lack of consensus on the most appropriate therapeutic approach for different stages of the disease because of limited number of patients.[2]

MATERIALS AND METHODS

We reviewed the case records of all children less than 12 years of age, diagnosed with testicular tumours between January 1995 and December 2010 in the Department of Pediatric surgery at the Children's Hospital of Tunis. We recorded age at presentation, pre- and postoperative images, individual treatment and outcomes for all the children. The tumour review criteria included presentation, radiological investigations, tumour markers, evidence of metastases, treatment modalities, histopathology and outcomes.

All the children who underwent testes preserving surgery were evaluated for subjective evidence of testicular atrophy. The cosmetic, sonographic, and recurrence-free outcome was evaluated at follow-up with a physical examination and ultrasonography.

RESULTS

A total of eight patients presenting with testicular tumour were included. The mean age of them was 3.5

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years (range three months –11 years). The presenting symptom was a painless testicular mass gradually increasing in size in all cases. Five lesions were in left side and three in right one [Figure 1].

Among the eight prepubertal children with testicular tumours, four (50%) had mature teratomas, one (12.5%) epidermoid cysts, one (12.5%) immature teratoma and two (25%) had yolk-sac tumours. Clinical manifestation was testicular swelling and pain.

Alpha-foetoprotein (AFP) was elevated in all the two (25%) children with yolk-sac tumours, and the levels were normal for age in the rest. The children with yolk-sac tumours underwent radical inguinal orchiectomy [Figure 2]. The clinical stage was I in one patient, and one patient had retroperitoneal metastasis (stage II) and was given adjuvant chemotherapy. All these children were under close follow-up ranging from 12 to 36 months. None of these children had any recurrence or postoperative metastasis in the retroperitoneum or in the lungs.

Overall six (75%) of the tumours were benign on histopathological examination. In four of these benign tumours, inguinal radical orchiectomy was done, and in two children testicular preserving surgery was performed. At follow-up period median of 36 months, all these children were free of recurrence, testicular atrophy, or persistent testicular pain.

DISCUSSION

It is traditionally known that paediatric testicular tumours are very rare and the majority of them are benign. In fact, there are important differences between testicular tumours in children and those occurring in adults. These differences involve tumour histopathology, malignant potential and metastasis.

The large majority of testicular tumours in adolescents and adults are malignant GCTs, most commonly mixed GCT with pure seminomas occurring in older men. In contrast, the most common GCT in children is benign teratoma and the most common malignant tumour in children is a yolk-sac tumour, which is very rare in its pure form in postpubertal patients.[3,4]

Taskinen reported a population of 34 patients with testicular tumour, aged 0.2-18.5 years with 26 prepubertal ones. In 11 of the 34 patients (32%), the tumour was malignant and in 23 (68%) benign. The final histological diagnosis of malignant tumour was: yolk-sac tumour in six (18%) and the most frequent benign tumours were teratoma (16 cases or 47%), Leydig-cell tumours in two cases (6%), and epidermoid cyst in two cases (6%). Benign tumours were more frequent in prepubertal than in adolescent patients. Twenty of 26 (77%) patients younger than 13 years had a benign tumour. In contrast, the tumour was benign in only three of the eight (38%) aged more than 13 years.[3]

In the study of Villegas of 27 testis tumours, there are 15 prepubertal and 12 postpubertal ones. The main clinical sign was painless testicular mass in 14 cases (52%). Benign lesions were seen in 17 cases (63%) and malignant in only 10 cases (37%). In prepubertal patients, benign lesions were present in 73% of cases (11/15).

In other way, Pohl and al studied the histopathological features of all paediatric testicular tumours removed at several major referral centres. They found that the incidence of malignant tumours in children was even less than that described in the Prepubertal Testis Tumor



Figure 1: Clinical aspect of left testicular tumour



Figure 2: Peroperative view of left testicular tumour and inguinal orchidectomy

Registry. Teratoma accounted for 48% of the tumours, with yolk-sac tumours accounting for only 15%. Overall, 74% of the tumours in that series were benign.[5]

Molecular studies have confirmed the distinct nature of prepubertal testicular tumour from those of adult. A chromosomal mutation in the short arm of chromosome 12p has been found in nearly all malignant adult GCTs. In contrast, in yolk-sac prepubertal tumour, there are no changes at chromosome 12p, but others changes exist in chromosomes 1, 6 and 20.[6]

The left testis was involved more than the right testis for both primary malignant neoplasms and benign neoplasms, which differs from that the data reported by Ciftci *et al.*, who found a right side preponderance (29:20, right/left) in testicular neoplasms.[2]

The majority (90%) of patients with testis tumours present with a scrotal mass noted by the patient, a parent, or a health care provider. These masses are typically hard and painless and must be distinguished from extratesticular masses.[2,4,7] A hydrocele is associated with a testicular tumour in 15–40% of cases, and a thorough examination is needed to determine the presence of other pathology, such as epididymitis, inguinal hernia, and testicular torsion. The latter might manifest as a painless mass in a neonate with little scrotal wall inflammation if it occurred prenatally. Acute abdominal pain could be the initial symptom of tumour in ectopic testis. These situations can cause misdiagnosis varying from 7 to 22%.[1,2,8] In many cases, tumour can be primary detected during operation (because of acute scrotum or persistent hydrocele).[7]

When the physical examination is equivocal, ultrasound is an excellent tool for distinguishing intra-testicular from extratesticular masses. Although ultrasound cannot reliably distinguish malignant from benign testicular tumours, cystic tumours are more likely to be benign.

The detection of testicular neoplasms with ultrasonography approaches 100%. Benign tumours tend to be well circumscribed with sharp borders and decreased blood flow on Doppler studies. Epidermoid cysts present as firm, well defined intratesticular lesions that, on ultrasound, have a central hypoechoic region or mixed echogenicity surrounded by an echogenic rim.

Yolk-sac tumours tend to be more solid in appearance, classically hypoechogenic and homogeneous, but can present with nonspecific signs. In cases where there is a suspicion of malignancy, cross-sectional imaging with CT or MRI of the abdomen, pelvis, and chest are needed to determine the preoperative stage and plan therapy, since around 20% of yolk-sac tumours are associated with lung metastases[2,8] [Figure 3].

Testis tumour markers are an important tool in the evaluation of testis tumours in children and adolescents. Both human chorionic gonadotropin (HCG) and AFP are important testis tumour serum markers. Although HCG is elaborated in case of malignant GCTs that were rare in prepubertal children, therefore, HCG is not a helpful marker in this population. AFP is elaborated by 90% of yolk-sac tumours in children and is more helpful in preoperative distinction between yolk sac and other tumours. But the level of AFP is normally high in healthy newborn and decrease progressively within six months age. So this marker is less helpful in young infants.[1,6,7]

A preoperative metastatic evaluation may be undertaken in patients older than six months who have elevated AFP levels and likely harbor yolk-sac tumours. Metastases from yolk-sac tumours typically occur in the lungs and retroperitoneal lymph nodes. A chest X-ray or computerized tomography (CT) scan, as well as an abdominal CT scan, should be obtained.[2]

The postoperative AFP levels should be followed. The half-life of AFP is approximately five days and failure of an elevated AFP to decline after removal of the primary tumour can be caused by a persistent metastatic disease.[1]

There is no universally accepted staging system for paediatric testis tumours. Prepubertal patients are

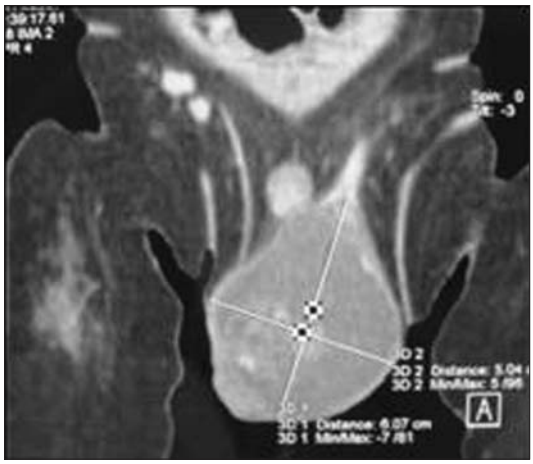


Figure 3: MRI of the pelvis showing tissular tumour of the left testis

generally staged based on the local extent of disease, the presence or absence of metastatic disease based on radiographic imaging (CT of the chest and abdomen), and the persistence or decline of elevated tumour markers postoperatively.[2]

Treatment depends on histological type. Classically, radical inguinal orchidectomy is the universally accepted initial surgical management of malignant GCTs. Scrotal orchidectomy should not be performed to avoid tumour seeding and scrotal violation.[2,7] For benign lesions, partial orchidectomy in patients under 12 years with normal AFP could be considered.[7]

Retroperitoneal lymph node (RPLN) excision is a controversial issue. Haematogenous spread is more frequent in children than in adult, and at presentation 70% of patients with malignant GCT were at stage I, and we were able to predict the extent of the disease by clinical and radiography. In addition, RPLN excision was found not to increase the survival rates in children and have major complications rates as high as 19%.

Therefore, RPLN excision for stage I disease is not indicated and RIO alone is suffusing for malignant GCTs. It should be preserved for patients how have persistent elevation of AFP after orchidectomy and for patients presenting with stage II and III disease with definitive abnormal CT scan.

Following excision of the primary tumour by inguinal incision with high ligation of the spermatic cord, benign tumours require no further evaluation or treatment. Treatment options for potentially malignant tumours include monitoring, chemotherapy, and radiation therapy. Virtually all GCTs are sensitive to platinum-based multiagent chemotherapy which plays a major role in their management. Radiation therapy is primarily used in treating seminoma which is a very rare tumour in the paediatric population. The specific adjuvant therapy for a given patient is dependent on tumour histology and stage.

CONCLUSIONS

Testicular tumours in children are rare. They occur most often as a scrotal mass which can sometimes be painful. During prepubertal period, they are dominated by benign lesions; malignant tumours represent only 1/4 of cases. Their diagnosis based on ultrasound, CT and dosage of APF. Treatment depends on histological type. Classically, radical inguinal orchidectomy is the universally accepted initial surgical management of malignant GCTs followed by chemotherapy, and a radiation if necessary. For benign lesions and if tumour markers were negative, testis-preserving surgery can be proposed.

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