Malignant Scrotal Masses in Children

Philip R. Exelby, M.D.

Cancer of the testis and paratesticular structures in children may be divided into three groups: embryonal adenocarcinoma of the prepubertal testis; teratocarcinoma of the testis; and paratesticular rhabdomyosarcoma. All are rare, together comprising less than 1.5 percent of all childhood cancers. The cause of testicular tumors in children is unknown. However, the rarity of these tumors in Negro and Asiatic children,1,2 the occasional occurrence of familial testicular cancer3,4 and the rare gonadoblastoma in dysgenetic gonads suggest genetic factors. There is an increased incidence of testicular cancer in patients with cryptorchidism according to Gilbert, but orchiopexy may not protect these children against the later development of cancer.5,6 Cancerous change in the cryptorchid rarely occurs in childhood although we have seen one infant and one adolescent whose cancer developed in an abdominal testis. The influence of hormones in the development of testicular cancer is uncertain. However, teratocarcinoma and paratesticular rhabdomyosarcoma show a marked rise in incidence at the onset of puberty and embryonal adenocarcinoma, seen largely in infancy, might be related to the effects of maternal estrogen.

The management of children with these tumors is controversial, but there is no doubt that early diagnosis and a better understanding of the behavior of the different testicular tumors will improve cure rates.

Histologic Types

Embryonal Adenocarcinoma

These tumors are described by a variety of terms such as embryonal carcinoma of infancy, orchioblastoma, endodermal sinus tumor, Teilum's tumor and adenocarcinoma with clear cells. Young believes that they are all histologic variants of the same tumor and may be properly classified as embryonal adenocarcinoma of childhood.7

Teratocarcinomas

These tumors contain tissue derived from all germ layers and, by definition, at least one malignant component, most commonly embryonal carcinoma. In our experience all children had embryonal carcinoma as one malignant component; in some cases, an additional malignant element was seen, such as chorionicarcinoma, seminoma and spindle cell sarcoma.

Paratesticular Rhabdomyosarcomas

Approximately 20 percent of embryonal rhabdomyosarcomas in children occur in the lower genitourinary tract, most commonly in bladder, vaginal and paratesticular tissue. Paratesticular lesions originate in the distal spermatic cord structures but the tissue of origin is uncertain. A primitive mesenchymal or teratomatous origin has been suggested.

Dr. Exelby is Chief, Pediatric Surgical Services, Memorial Hospital for Cancer and Allied Diseases, New York, New York.
Figure 1. Clinical Presentation

(a) Embryonal adenocarcinoma, teratocarcinoma arising in scrotal testis
(b) Teratocarcinoma following orchiopexy
(c) Empty left scrotum, tumor arising in abdominal testis
(d) Paratesticular rhabdomyosarcoma, distal cord
Age Incidence

Embryonal adenocarcinoma is largely a disease of infancy. Two-thirds of patients will be under two years of age at diagnosis. The disease is rare in children over four years old. Teratocarcinoma is generally seen in postpubertal boys and is uncommon in children under fourteen years old. Paratesticular rhabdomyosarcoma shows two peaks of incidence, one in early childhood and the second in adolescence. In our experience approximately 50 percent of tumors arise between the ages of 13 and 16 years.

Clinical Presentation

These tumors commonly present as nontender, slow-growing scrotal masses. (Fig. 1.) Occasionally, the patient will have associated pain usually related to a complication such as torsion. Rarely, abdominal swelling in an undescended testis with an empty scrotum is the presenting symptom. Paratesticular rhabdomyosarcomas initially present as swellings in the upper part of the scrotum or the distal spermatic cord. They tend to grow into the testis and become indistinguishable from primary testicular cancer. Approximately 25 percent of all malignant tumors of the testis in children will have an associated hydrocele which can mask the testicular mass. Examination of the inguinal region may show a previous herniorrhaphy or orchiopexy scar. Rarely, a teratocarcinoma secreting chorionic gonadotrophin may produce precocious puberty as the presenting symptom.

Course of Disease

Although these three tumors present initially as scrotal masses and are largely indistinguishable clinically, their course is quite different. Embryonal adenocarcinomas grow slowly and metastasize primarily to the lungs. (Fig. 2.) Although there are isolated reports of metastasis to para-aortic nodes, it has never occurred in our experience. Certainly the most common mode of spread seems to be hematogenous. Teratocarcinomas in children spread to lymph nodes in the

Fig. 2: Embryonal adenocarcinoma of infancy; usually negative lymph nodes.
same manner as adult testicular cancer; namely, to the high ipsilateral para-aortic nodes. (Fig. 3.) In our experience approximately 50 percent of these children will have positive high para-aortic nodes at the time of initial presentation. Simultaneous or subsequent metastasis to the lungs may also be seen. Paratesticular rhabdomyosarcomas which initially form a mass in the distal cord spread locally to involve the testis and become indistinguishable from a primary testicular cancer. Their metastatic spread is different from the previous tumors in that it tends to involve the iliac nodes and the para-aortic lymph node chain. (Fig. 4.) In our experience approximately 50 percent of patients had involvement of the common iliac nodes, and 65 percent had involvement of para-aortic nodes at the time of diagnosis. Hematogenous spread follows nodal involvement and is seen commonly in the lungs and bones.

**Diagnosis**

Early diagnosis is very important in this group of diseases, which will be fatal to the child within two years if improperly treated. In our experience an average delay of three months was noted between onset of symptoms and definitive diagnosis. An inguinal biopsy should be performed with the cord clamped. If the diagnosis is cancer, a radical orchiectomy should be immediately performed. Subsequent management then depends on the histologic type and clinical behavior of the tumor.

In the infant, embryonal adenocarcinoma is adequately treated by radical orchiectomy with high ligation of the cord. There is no evidence that retroperitoneal node dissection, radiation therapy or prophylactic chemotherapy improves survival in this disease. In the small number of children who have lung metastases at the time of diagnosis, radical orchiectomy is followed by four drug cyclic chemotherapy. (Fig. 5.) Patients with teratocarcinoma, which spreads to ipsilateral para-aortic nodes, should be treated by ipsilateral paracaval or
paraortic node dissection in addition to orchiectomy. Those children with paratesticular rhabdomyosarcomas, which are more aggressive locally and spread to the iliac and para-aortic nodes, are currently managed by radical orchiectomy, hemiscrotectomy, external iliac, common iliac and retroperitoneal node dissection. Radiation therapy is given to the retroperitoneal area in patients with teratocarcinoma and positive nodes. In patients with paratesticular rhabdomyosarcoma, radiation therapy to the whole retroperitoneal node area, hemipelvis and inguinal region is given if margins of resection are positive for cancer or if retroperitoneal or iliac nodes show metastases. Four drug cyclic chemotherapy is recommended for all children with teratocarcinoma or paratesticular rhabdomyosarcoma regardless of the stage of the disease. (Fig. 5.) Drug therapy should then be continued for two years, which appears to be the risk period for local recurrence or metastasis.

Results of Treatment

In our experience the overall survival of children with embryonal adeno-carcinoma is 75 percent. Three children had lung metastases at the time of diagnosis and died of their disease. Six patients with stage A disease were treated by orchiectomy alone and five survived. Ten children had an orchiectomy followed by retroperitoneal node dissection. None of the excised nodes showed metastatic disease. Therefore, it appears that retroperitoneal node dissection added little to survival and is no longer recommended as part of surgical management. In the teratocarcinoma group, two patients were treated by orchiectomy alone and one, an infant, survived. Twelve patients were treated by orchiectomy followed by retroperitoneal node dissection, and 50 percent of these children are surviving free of disease after more than two years. In the children with paratesticular rhabdomyosarcoma the disease was advanced at the time of

Fig. 4. Paratesticular rhabdomyosarcomas metastasize to iliac and paracaval nodes
diagnosis. Two-thirds of these children had retroperitoneal nodes positive for cancer, and all of these children eventually died of advanced metastatic disease. Prior to 1960 the survival rate was zero; between 1960 and 1970 the cure rate improved to almost 50 percent. With current management including radical surgery, selective radiation therapy and aggressive chemotherapy, the expected survival is now 75 percent.

Factors Affecting Survival

(1) Early diagnosis. As with most other cancers early diagnosis has a marked influence on the cure rate. In childhood cancers, which are more rapidly progressive than adult cancers, it is imperative that diagnosis is early and appropriate treatment is instituted immediately. (2) Age at diagnosis. Tumors occurring in infancy such as embryonal adenocarcinomas and paratesticular rhabdomyosarcomas in the young patient have a much better prognosis than those which develop in older children. Until 1970 all survivors of paratesticular rhabdomyosarcoma were under the age of seven years. With improvement in management, especially chemotherapy, the prognosis in older children has markedly improved in recent years.

(3) Combined treatment. Greater understanding of the natural history of these diseases and the use of combined treatment—surgery, radiation therapy—are producing further improvements in cure rates.

References

1. Fergusson, J. D.: Tumours of the testis. Brit. J. Urol. 34: 407-421, 1962.
2. Li, F. P., and Fraumeni, J. F., Jr.: Testicular cancers in children: epidemiologic characteristics. J. Nat. Cancer Inst. 48: 1575-1581, 1972.
3. Pugh, R. C.: Tumours of the testis. Frequency and race. In: Raven, R. W. (ed.), Cancer, London, 1958. Vol. 2, pp. 241-248.
4. Tilman, A. J.: The racial incidence of testicular tumours. S. Afr. Med. J. 43: 97-98, 1969.
5. Collins, D. H., and Pugh, R. C.: Classification and frequency of testicular tumours. Brit. J. Urol. 36: Suppl. 1-11, 1964.
6. Hutter, A. M., Jr.; Lynch, J. J., and Shnider, B. L.: Malignant testicular tumours in brothers. JAMA 199: 1009-1010, 1967.
7. Gilbert, J. B., and Hamilton, J. B.: Studies in malignant testis tumors. III. Incidence and nature of tumors in ectopic testes. Surg. Gynecol. Obstet. 71: 731-743, 1940.
8. Gilbert, J. B.: Studies in malignant testis tumors: tumors developing after orchidopexy: report of 2 cases and review of 63. J. Urol. 46: 740-747, 1941.
9. Young, P. G., et al.: Embryonal adenocarcinoma in the prepubertal testis. A clinicopathologic study of 18 cases. Cancer 36: 1065-1075, 1970.
10. Whitmore, W. F., Jr.: Germinal tumors of the testis. Proc. Nat. Cancer Conf. 6, 1970. Pp. 219-245.