TERATOMAS IN PEDIATRIC AGE GROUP: EXPERIENCE WITH 75 CASES

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ABSTRACT

The clinicopathological features of 75 children under the age of 12 years with teratomas are reviewed. Tumors arose in the following anatomic sites: sacrococcygeum (n=49), ovary (n=10), Testis (n=5), oral cavity (n=3), retroperitoneum (n=2) and others (n=6). Fifty five (74%) presented within the first year of life. Excluding the gonadal tumors, male-female ratio was 2 : 5. Majority of the tumors had only mature tissues. Such patients and those 9 patients in whom the histology was not specified, underwent excision alone and had 95% early survival rates. Five patients had admixture of mature and immature tumors. Nine patients had malignant tissues. Germ cell tumors containing only malignant component, but no mature or immature teratomatous tissues were excluded from the series. The patients with immature and malignant tissues underwent multimodal therapy including surgical excision, multiagent chemotherapy (VAC regimen) and at times radiotherapy. Mortality in patients with immature and malignant teratomas was 20 and 66.7%, respectively. Besides histology, the only factor which affected prognosis, especially in case of sacrococcygeal teratomas was the age at the time of presentation. Our experience highlights the importance of early recognition and complete surgical excision of teratomas in the pediatric age group

Key words: *Teratoma, Mature teratoma, Germ Cell tumor.*

Teratomas are notable for their diversity in anatomic location and biologic behavior. Through this report of 75 patients treated over 22 years, we wish to review the clinicopathological features of teratomas in the pediatric age group and also attempt to identify the factors that influence therapy and prognosis.

Material and Methods

Seventy five children under the age of 12 years were treated at the Department of Pediatric Surgery, SMS Medical College and attached SPMCHI during a 22 year period (October 1970 to September 1992). The clinical records of the patients were reviewed and follow-up data on all but eleven were obtained. The teratomas of the various anatomic locations were classified in three pathologic categories based on histologic findings (*i*) mature teratomas, (*ii*) immature teratomas (containing embryonic with or without mature tissues), and (*iii*) malignant teratomas. The malignant tumors having no immature and/or mature tissues were excluded from this series. In 9 cases the histology in regard to the pathological category was not specified in biopsy reports.

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Received for publication: February, 9, 1992; Accepted : February 3, 1993

Results

Fifty five of these patients presented within the first year of life. There was a distinct female preponderance (*Table I*). Fifty two patients had mature tumors, 5 had immature tumors and 9 patients had malignant teratomas (*Table II*). Of the 75 patients, 11 patients have been lost to follow-up. Another 10 children, 6 females and 4 males, died either of malignancy or post-operative complications. Fifty four patients are living without known residual tumors; 46 have been followed up for more than five years. The survivors include three patients of immature teratomas (one each of sacrococygeum, testis and ovary). One child with malignant sacrococcygeal teratoma is also alive and well without metastasis or local recurrence one year after the surgery.

Considering the wide biological diversity of teratomas in regard to the site of origin, they have been discussed separately briefly below.

Sacrococcygeal Teratomas: Sacrococcygeal tumors were the most frequent lesions observed in 49 (65.3%) of the total patients (*Table I*). The majority of these tumors had only mature tissues. Malignancy was seen in 6 patients; 5 patients were in Stage III and one was in Stage IV(1). We did not come across any patient initially labelled as benign(mature) lesion recurring as malignancy later in the series. Majority of the cases (Altman's type I and II)(2) represented with an obvious mass in the region. Others (Altman's type III and IV) presented with features of urinary and bowel obstruction. According to Altman's classification, 33 (67.3%) patients were type I; types II, III and IV constituted 16.3, 6.1 and 10.2%, respectively. The tumors ranged from 2 cm to 25 cm, with most lesions having an admixture of solid and cystic components. Altman's III and IV tumors presented late, usually after 6 months of age and as many as half of them were malignant, having embryonal cell carcinoma component (*Table II*).

Benign lesions were treated with complete excision; there were 2 post-operative deaths. Seven out of the eight patients with immature and malignant teratomas received multiagent chemotherapy. Regimen includes monthly cycles of vincristin (1.5 mg/m^2 IV day 1 and 7), actinomycin-D (0.015 mg/kg/day IV day 1 to 5) and cyclophosphamide (500 mg/m^2 IV day 1 to 3). One patient having extensive metastasis at initial presentation refused any adjuvant therapy. The malignant component in all the 6 cases was embryonal cell carcinoma and all except one (mentioned above) succumbed to widespread metastasis within months of initial diagnosis.

Ovarian Teratomas: The ovary was second only to sacrococcygeal area as the commonest site for occurrence of teratomas. There were 10 cases seen, usually presenting between 5 to 10 years of age. The youngest child was 3 years old. The usual presentation was abdominal pain and pelvic/abdominal mass. One patient presented as acute abdomen following torsion of fallopian tube and ovary. Calcification was seen in 40 cases of *X*-ray examination. Unilateral salpingo-oophorectomy was done in all patients, none of them had peritoneal seedlings or bilateral involvement at initial presentation. Histology was specified in 9 cases. Seven had mature tumors, immature and malignant tumor were seen in one patient each. Patients with immature and malignant tumors received adjuvant chemotherapy. The patient with malignant teratoma succumbed to widespread metastasis.

TABLE I–*Anatomic Site and Sex Distribution of Teratomas*

Site	No. (%)	Male	Female
Sacrococcygeal	49 (65.3)	12	37
Ovarian	10 (13.3)	-	10
Testicular	5 (6.7)	5	-
Oral Cavity	3 (4.0)	1	2
Retroperitoneal	2 (2.7)	-	2
Cervical	2 (2.7)	2	-
Nasopharyngeal	1 (1.3)	-	1
Lumbosacral	1 (1.3)	-	1
Perineal	1 (1.3)	1	-
Gastric	1 (1.3)	1	-
Total	75	22 (29%)	53 (71%)

TABLE II–*Correlation of Age at Presentation and Histopathology of Teratoma*

Site	Type	Age at diagnosis (mo)				Total
		0-2	2-6	6-12	>12	
Sacrococcygeal	Mature	21	13	1	1	36
	Immature	1	1	-	-	2
	Malignant	-	-	4	2	6
	Not specified	2	2	1	-	5
Ovarian	Mature	-	-	-	7	7
	Immature	-	-	-	1	1
	Malignant				1	1
	Not specified	-	-	-	1	1
Testicular	Mature	-	-	-	2	2
	Immature	-	-	-	1	1
	Malignant	-	-	-	2	2
Oral Cavity	Mature	1	-	-	1	2
	Not Specified	1	-	-	-	1
Retroperitoneal	Not specified	-	1	-	1	2
Cervical	Mature	1	-	-	-	1
	Immature	1	-	-	-	1
Nasopharyngeal	Mature	1	-	-	-	1
Lumbosacral	Mature	1	-	-	-	1
Perineal	Mature	1	-	-	-	1
Gastric	Mature	1	-	-	-	1
	Total	32	17	6	20	75

Testicular Teratomas: Five patients had testicular teratomas, two on the right and three on the left side. The youngest patient was 1½ years of age and the oldest 5 years at the time of diagnosis. All of them presented with painless enlargement of testis. High orchidectomy was performed in all the patients. The two patients with malignant teratomas (one seminoma and another embryonal cell carcinoma)and one child with immature teratoma received adjuvant chemotherapy. Both the boys with malignant testicular tumors had normal retroperitoneal imaging pre-operatively and were not subjected to retroperitioneal lymph node dissection. One child with malignant tumors died of metastasis and another was lost to follow-up.

Oral Teratomas: Two male neonates and one six-year old female child presented with teratomas arising in the oral cavity. Both boys came with respiratory distress, whereas the girl presented with a large mass arising from inside of the right cheek. All three had complete excision. The details of these have been reported previously(3).

Retroperitoneal Teratomas: Two patients presented with asymptomatic retroperitioneal mases causing no bowel or urinary complaints. Inravenous pyelograms proved the masses of non-renal origin. There was evidence of clacification in both tumors. Complete excision was possible in both the cases; there were not enlarged retroperitoneal lymph nodes or peritioneal seedings.

Cervical Teratomas: Two male neonates presented with cervical teratoma in first few days of life. Both presented with anterior neck masses which showed evidence of calcification on *X*-rays. One of them had severe respiratory distress at presentation. Complete excision was easily done in both cases; however, the latter child died in the early post-operative period.

Nasopharyngeal Teratoma: We encountered an unusual nasopharyngeal teratoma in a male neonate which was attached to basisphenoid as well as the mandible, thus allowing incomplete opening of mouth and mild respiratory distress. Complete excision was successfuly done.

Lumbosacral Teratoma: One female neonate was admitted soon after birth with swelling in the lumbosacral region. The patient was operated for meningomyelocele. Coincidental teratoma discovered intraoperatively was also excised. The patient died in immediate post-operative period.

Perineal Teratoma: One male neonate was seen with a perineal teratoma simulating appearance of male external genitalia. The details have been previously published(4).

Gastric Teratoma: A 6-month-old male child presented with intermittent post parandial vomiting. Examination revealed a 5 × 5 cm firm, mobile, non-tender mass in the epigastric region. *X*-ray showed irregular clacification in the region of the mass. Intravenous pyelography was normal and barium enema showed downward displacement of transverse colon. Laparotomy revealed a mass of 8 × 8 × 6 cm arising from the posterior wall of the stomach adjacent to the greater curvature. Total excision of the tumor was done and the defect in the stomach repaired. The post operative period was uneventful.

Discussion

Teratomas are the most diverse group

of germ cell tumors seen in childhood accounting for 3% of all childhood malignancies(5). Sacrococcygeum and the gonads were noted to be the most common sites, similar to previous experience of others(6-10). Another feature common to most series is a female preponderance, accounting for 71% cases in our series. Majority of the patients had only mature tissues in the teratoma. We did not come across a single case of recurrence at the site of surgery or later transformation into malignancy. This could be related to our policy of complete excision. For example, the sacrococcygeal teratoma excision always included excision of coccyx. Two of the three deaths in this group occurred in the post-operative period due to secondary hemmorrhage and aspiration pneumonitis. The third patient died of associated cardiac anomaly. The excellent results in the case of mature tumors have been reported earlier also(9).

The results in the patients with immature teratomas were comparable with those having exclusively mature tissues. Such a benign course of these tumors has been reported earlier also(7). The role of adjuvant chemotherapy has not yet been confirmed in cases of immature teratomas and some authors feel that if surgery has been adequate, chemotherapy may be withheld, but a careful follow-up with tumor-markers (alfa-fetoprotein, *etc.*) monitoring is mandatory(11). But a few series have reported a significantly lower survival with immature tumors as compared to the mature tumors and recommend adjuvant chemotherapy and/or radiotherapy especially in tumors of ovary, retroperitoneum and central nervous system(9). This is particularly so if a complete resection is not possible.

The overall incidence of malignancy was 13%, lower than that quoted by other series(10). The tumors included 7 cases of embryonal cell carcinoma and 2 cases of germinoma (one each of seminoma testis and dysgerminoma ovary). The mortality in this subgroup of teratomas was as high as 66.7%. Majority of the patients having embryonal cell carcinoma did not survive. This group was characterised by low surgical resectability related mainly to the anatomic site, large size of the tumor and invasion of adjacent tissues or organs. Recurrence at the local site as well as metastasis to the lungs, liver and peritoneum were noted. The patient with malignant ovarian teratoma also died within 4 months of salpingo-oophorectomy. The spread of malignancy in ovarian teratomas is via peritoneal fluid rather than chance of cure(12). In such situations, the role of adjuvant therapy is vital. Billmire and Grosfeld(10) reported a 100% survival in resectable malignant teratomas in which adjuvant therapy was also used as compared to 17% in patients undergoing surgery alone. However, the most important variable affecting long-term survival is the surgeon's ability to completely resect the tumor. The same authors(10) have quoted a dismal 11% survival in patients who had unresectable tumors at diagnosis and were subjected to chemotherapy followed by second-look operation. Favorable results have been reported for the malignant testicular teratomas as compared to those of extra-testicular sites(7,10).

Regarding the prognostic indicators, the value of various factors including the size of tumor, cystic or solid character, presence of calcification has been assessed by several authors(2-7). The histology undoubtedly affects the prognosis. We studied the risk factor of developing malignancy as a related to the age of presenta-

tion and found that age at diagnosis was of no predictive value as regard to the teratomas of the sites other than sacrococcygeal region. But in regard to sacrococcygeal tumors, the incidence of malignancy rose from nil under 6 months of age to 50% in those who were diagnosed after this age. In Altman's survey of 405 patients, malignancy was noted in 7% for neonatal tumors, 37% at 1 year of age and 50% at the age of 2 years(2). Billmire and Grosfeld(10) have questioned the suggestion of previous reports that malignant transformr time and feel that the percentages in Altman's survey are rather overquoted.

Our experience highlights the importance of early recognition and complete surgical excision of teratomas in the pediatric age group. Prognosis in case of malignant teratomas is quite dismal and probably more aggressive chemotherapeutic regimens should be tried.

REFERENCES

1. Schropp KP, Lobe TE, Rao B, *et al.* Sacrococcygeal teratoma: The Experience of four decades. J Pediatr Surg 1992, 27: 1075-1079.
2. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey, 1973. J Pediatric Surg 1974, 9:389-398.
3. Sharma S, Gupta AK, Sharma AK, Sogany K. Oral teratomas in Pediatric age group. Santokba Durlabhji Memorial Journal 1987, 11: 161-163.
4. Agarwal LD, Prabhakar G, Sharma K, Shukla A, Sharma CS, Sharma AK. Perineal teratoma: A case report. Santokba Durlabhji Memorial Hospital Journal 1991, 14: 361:362.
5. Brodeur GM, Howrath CB, Pratt CB. Malignant germ cell tumors in 57 children and adolescents. Cancer 1981, 48: 1980-1898.
6. Berry CL, Keeling J, Hilton C. Teratoma in infancy and childhood: A review of 91 cases. Cancer 1969, 98: 241-252.
7. Carney JA, Thampson DP, Johnson CL, Lynn HB. Teratomas in children: Clinical and Pathologic aspects. J Pediatr Surg 1972, 7: 271-282.
8. Mahou GH, Woolley MM, Trivedi SN, Landing BH. Teratomas in infancy and Childhood: Experience with 81 cases. Surgery 1974, 76: 309-318.
9. Tapper D, Lack EC. Teratomas in Infancy and Childhood: A 54-year experience at the Children's Hospital Medical Centre. Ann Sug 1983, 198: 398-410.
10. Billmire DF, Grosfeld JL. Teratoma in childhood: Analysis of 142 cases. J Pediatr Surg 1986, 21: 548-551.
11. Ein SH, Mancer K, Adeyemi SD. Malignantsacrococcygeal teratoma–endodermal sinus, yolk sac tumor in infants and children: A 32-year review. J Pediatr Surg 1985, 20: 473-477.
12. Kosloske AM, Favara BE, Hays ET. Management of immature teratoma of the ovary in children by conservative resection and chemotherapy. J Pediatr Surg 1976, 11: 839-844.