
Congenital mesoblastic nephroma is the only primary renal neoplasm with radiographically demonstrable function. Renal function within two typical congenital mesoblastic nephromas was demonstrated by computed tomography in one case and renal scintigraphy in the other. Excretion of contrast medium and radiotracer was due to functioning nephrons trapped within the tumor.—Prem Puri


This work stems from the efforts of the authors as representatives of the International Conferences of Neuroblastoma Staging and Response Criteria to standardize, diagnosis, staging, and response criteria. Without reproducing the results of the authors work here, I would urge every pediatric surgeon who treats neuroblastoma, which accounts for 8% of all pediatric cancers and strikes nine children less than 15 years of age per million yearly in the United States, to review this report. The New International Neuroblastoma Staging System, the extent of disease work-up recommendations, and definitions of response to treatment that have been put forth by this international cooperative effort will help to make comparisons between treatment regimens more uniform and thereby greatly facilitate the world-wide management of this all-too-common pediatric cancer.—Michael P. Hirsh and Ian T. Cohen


This review of 26 patients with neuroblastoma from Children's Hospital of Philadelphia analyzes the use of the I-131 metaiodobenzylguanidine nuclear medicine scan (MBG) which has been useful in detection of neuroectodermal tumors. Thirty-eight scans were obtained in these patients all of whom had been previously treated for their disease. Although the scan did seem to have some usefulness, it had a significant false negative rate in cases of anaplastic, very mature, or metastatic forms of this disease. This may be related to the pretreatment affecting uptake. The authors conclude that they can not recommend MBG scanning as replacement for other studies evaluating reoccurrence of neuroblastoma.—Michael P. Hirsh and Ian T. Cohen


The authors present two cases of mediastinal tumors (neuroblastoma, ganglioneuroma) which extended into the intraspinal canal. Incidence and symptomatology are discussed. It is mandatory to examine the intraspinal situation by means of CT combined with metrizamide myelography in all cases of paravertebral tumors. Simultaneous laminectomy and tumor removal in one operation is recommended in intraspinal extension of mediastinal tumors.—Thomas A. Angerpointner


Several authors have reported an association between neuroblastoma and congenital heart disease. Others contend that, unlike specific well-known associations between malignancy and congenital defects (Wilms' tumor and aniridia, leukemia, and Down's syndrome), no real relationship exists. The authors present three cases of cyanotic congenital heart disease in which subclinical neuroblastoma was found. The authors speculate that abnormal neural crest cell migration and development may be a common link between cardiac malformations and congenital neuroblastoma.—Prem Puri


Neuroblastoma and ganglioneuroblastoma have been associated with adrenal gland abnormalities. Cushing's syndrome and adrenocortical hyperplasia are among the reported diagnoses. The authors present a boy with salt-wasting adrenogenital syndrome (21-hydroxylase deficiency) who responded to cortisone acetate and intramuscular DOCA therapy and who later developed stage IV neuroblastoma with a left suprarenal mass. Eight reported cases of children with other adrenocortical abnormalities and neuroblastoma are reviewed. The authors suggest serial monitoring of urinary vanillylmandelic acid and homovanillic acid levels in children with congenital adrenal abnormalities.—Jeffrey L. Zitsman


One hundred sixty-two patients had tumors of high-grade osteosarcoma arising in the long bones of the extremity, of which the median age was 20 years (range, 3 to 75 years). There were 105 males and 57 females. One hundred seven patients developed recurrent disease with the lung as the sole site of metastases in 77 patients (72%), the lung and one additional site in 17 patients (16%), and an isolated site distant from the primary site in 13 patients (12%). Neither the location of the primary tumor nor the procedure performed on the primary tumor had any effect on the distribution of metastatic disease. The pattern of metastatic disease for patients who received adjuvant chemotherapy was similar to those who received none postoperatively. Of the 77 patients with lung metastases, 51 underwent resection of the metastases. Thirty-three of these (17%) are long-term disease-free survivors. Patients with metastases to the lung and other sites and patients with metastases to sites other than lung have a poor prognosis with only one long-term disease-free survivor. Thoracotomy has a limited role in the treatment of metastatic disease. Improvements in aggressive systemic chemotherapy are essential for improving survival in these patients.—George A. Rowe


This retrospective review uses a multivariate model to identify the factors that are predictive of recurrence and morbidity in children with differentiated thyroid carcinoma. The mean age at diagnosis was 13.3 years for 93 patients in a 35 year period. Seventy-one percent of these patients had nodal metastases. There were no deaths from thyroid cancer. In this group of children variables with significant predictive effect included age at diagnosis, type of thyroid surgery, type of lymphatic dissection, and histologic subtype. Younger patients (<11 years old) had a higher risk of major postoperative complications as well as an increased risk of tumor recurrence. The use of subtotal or total thyroidectomy increased the probability of a major complication compared with lobectomy or...
biopsy. Minor morbidity (bleeding, edema, transient hypocalcemia, etc.) was associated with radical neck dissections. The authors found that subtotal or total thyroidectomy, as well as radical neck dissection, should be avoided in children with differentiated thyroid carcinoma.—Tom Tracy, Jr


One quarter of the 106 patients with cystosarcoma phylloides presented at under 19 years of age. Thirty-four percent of the entire group waited for more than six months before seeking treatment. Large malignant tumors were found in three older patients. Six borderline tumors were found, also in women >30 years of age. Benign tumors were easily excised except in patients in which the tumor had replaced the whole breast necessitating mastectomy. Seventy-two percent of the tumors were <5 cm in diameter and unsuspected in the younger age group. Enucleation of these benign tumors resulted in 16% recurrence, and the presence of tumor free margins is stressed within a clear and detailed algorithm.—Tom Tracy, Jr


A female weighing 2,500 g with an in utero ultrasound diagnosis of an 8 cm hemangiopericytoma is reported. The tumor involved the lower lip and was partially removed in early infancy. The remaining tumor regressed spontaneously, leaving only a small scar at 20 months without additional therapy. The authors advocate close follow-up but avoidance of radical resection for this lesion.—Eugene S. Wiener


This is a case report of an 11-year-old boy who presented with respiratory distress and a large tumor involving the left chest wall and pleura. There was temporary improvement with chemotherapy and radiation therapy, although the patient died seven months after initial presentation. The pathology revealed malignant pleural mesothelioma, which generally results in death within 18 months of initial presentation. The authors discuss the importance of early diagnosis according to a literature review by the authors.—Eugene S. Wiener


The authors describe a 14-year-old boy with a 1-year history of increasing abdominal distention who was found to have a large multicystic abdominal mass, primarily about the hepatic flexure and the ileum. Most of the mass was removed. The findings were those of benign cystic mesothelioma which is differentiated from cystic lymphangiomata by the absence of smooth muscle in the wall of the cyst on light microscopy and the presence of cytokeratin in the lining cells and vimentin in the submesothelial cells in cystic mesothelioma. In addition, electron microscopy identifies laminal microvilli, many mitochondria, desmosomes, and intermediate filaments in cystic mesothelioma that are not present in cystic lymphangiomata.—Eugene S. Wiener


This important study from the National Cancer Institute includes adult patients, but addresses an often asked question that pediatric surgeons following patients with intraabdominal disease from Non-Hodgkin’s lymphoma (NHL) are faced with: Do residual masses post-therapy need to be surgically explored?

Two hundred forty-one patients over a 10-year period (1977 to 1986) with NHL were reviewed. Seventy-two (30%) had intraabdominal disease, and of these 29 (40%) had residual masses at the time of remission. Twenty-two of the 29 underwent reexploration and 21 patients (95%) had no residual disease. The 21 with no residual disease also have not relapsed.

Based on this information, seven patients of the 29 with residual masses were observed without reexploration. Five of seven are disease free in follow-up; two relapsed. Residual mass size and original tumor size did not correlate with residual disease, and the great majority of the residual masses had no viable tumor cells. Restaging laparotomy, therefore, has a low yield and the authors conclude that sizable residual abdominal masses in post-chemotherapy treated patients in remission can be followed clinically without surgical exploration.—Michael P. Hirsh and Ian T. Cohen


This study from the National Cancer Institute and a number of other oncologic centers reviews the educational achievement of a cohort of 2,283 long-term survivors of childhood and adolescent cancers. With the increased survival rates seen with childhood cancers in general, many concerns were raised about the quality of life of these survivors. If prolonged hospitalization and time away from school, or if the cancer or treatment itself cause physical or mental impairments to learning, or if the emotional drain on the patients’ families created an atmosphere non conducive to learning, then educational achievement and quality of life may be significantly less for childhood cancer survivors.

The 2,283 survivors were, therefore, surveyed and compared with 3,270 sibling controls. Fifteen percent of the survivors had CNS tumors, and these patients did have a significant educational deficit, particularly if the tumors were of ventricles or cerebral hemispheres, if surgery alone could not be used for treatment, or if the tumor was diagnosed at an early age. For the remaining 85% of survivors with non-CNS tumors, no educational deficits were seen. This should prove reassuring to the practitioners and families of survivors of childhood cancers.—Michael P. Hirsh and Ian T. Cohen