8. Rodrigues-Soriano J, Vallo A, Castillo G, Oliveros M. Natural history of primary distal renal tubular acidosis treated since infancy. J Pediatr 1982, 101: 669-676.

9. Palmer H, Cornfeld D. Primary renal tubular acidosis recognized and treated at 3 days of age. Clin Pediatr 1973, 12: 140-144.

10. Barratt TM. Urolithiasis and nephrocalcinosis. *In:* Pediatric Nephrology. Eds Holiday MA, Barratt TM, Vernier RL. Baltimore, William and Wilkins, 1987, pp 700-708.

11. Bettinelli A, Bianchetti MG, Giradin E, *et al.* Use of calcium excretion values to distinguish two forms of primary renal tubular hypokalemic alkalosis: Bartter and Bitelman Syndromes. J Pediatr 1992, 120: 38-43.

Pheochromocytoma

S.N. Tomaraei
M. Singh
A. Dhawan

Pheochromocytoma, rightly named as "pharmacologic bomb", is a rare tumor in the pediatric age group. It is one of the curable causes of hypertension in children. A review of Indian literature reveals only 6 children with pheochromocytoma reported so far(1-4).

The purpose of this communication is to report a cause of pheochromocytoma who presented with hypertensive encephalopathy, and to review the Indian literature with special reference to the mode of presentation of pheochromocytoma and to highlight certain points in its diagnosis and management.

From the Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

Reprint requests: Dr. Meenu Singh, Assistant Professor, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

Received for publication: January 16, 1992; Accepted: December 1, 1992

Case Report

A 9½-year-old male child was admitted to Nehru Hospital, PGIMER, Chandigarh with a history of convulsions few hours prior to admission. There was a history of palpitation, headache, abdominal pain and constipation for the last few months prior to admission. On admission he had sustained hypertension of 180/136 to 196/140 mm Hg. Systemic examination including fundus examination revealed no abnormality.

Intravenous pyelography (IVP) showed a right adrenal mass, suggestive of pheochromocytoma. Ultrasound scan of the abdomen revealed a well-defined mass in suprarenal region, measuring 3.3 × 2.6 × 3.1 cm, and CT scan abdomen showed a well-defined enhancing mass at the upper pole of the right kidney (*Fig. 1*). It measured 4 × 3 × 3.5 cm in size. Hemoglobin ranged from 14.5 to 12.5 g/dl and PCV 45 to 35% (after repeated plasma transfusions in order to maintain intravascular volume). Urine vannillylmandelic acid (VMA) estimations, on 3 different occasions, were 6.5 mg/24 h, 6.7 mg/24 h and 6.5 mg/24 h, respectively (normal

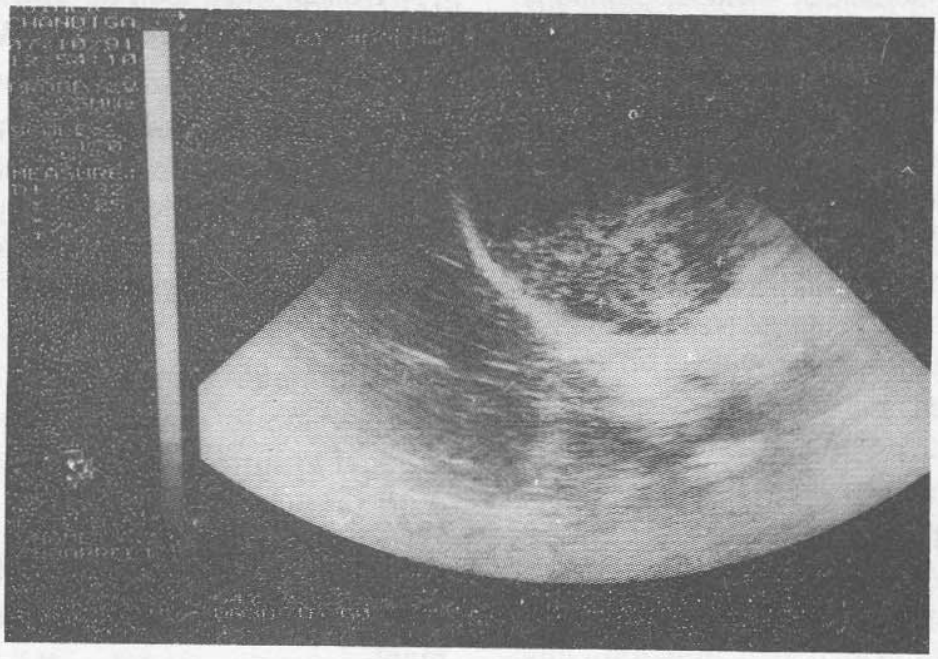

Fig. 1(a). Ultrasound and CT scan abdomen.

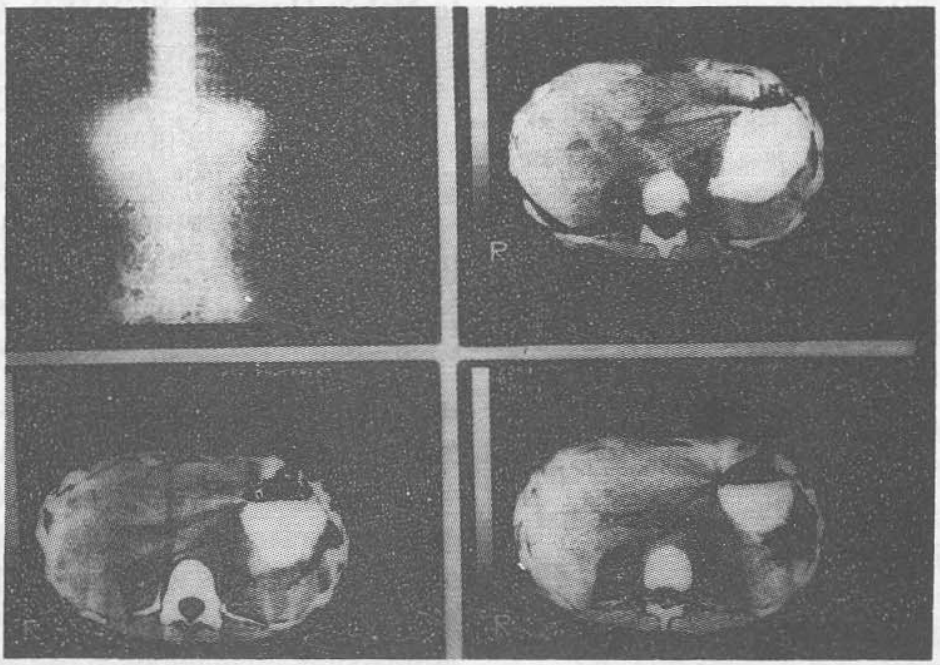

Fig. 1(b) Showing right-sided suprarenal tumor (+, tumor).

= 1-7 mg/24 h). Plasma catecholamines were elevated; epinephrine 1.8 ng/ml (normal = 0.41 ng/ml) and norepinephrine 3.4 ng/ml (normal = 1.6 mg/ml). ECG showed tachycardia and left axis deviation, and echocardiography revealed mild concentric left ventricular hypertrophy. Serum electrolytes, renal function tests, urine (routine

examination, microscopy, culture and prophobilinogen), serum calcium, phosphorus, alkaline phosphatase, chest *X*-ray and skull *X*-rays were all normal.

A diagnosis of pheochromocytoma was made. Initially he was managed with intravenous infusion of sodium nitroprusside and other antihypertensive drugs. Later, he was started on phenoxybenzamine (dibenzyline) 3 weeks prior to operation. Propranolol was subsequently added to control the tachycardia. He received plasma transfusions in order to maintain the intravascular volume. Blood pressure decreased to normal and the child was successfully operated. Histopathological examination of the right adrenal tumor showed features of pheochromocytoma. There was no evidence of malignancy. The postoperative period was uneventful. Investigations on follow-up visits showed: plasma norepinephrine 1.4 ng/ml (normal = 1.6 ng/ml), epinephrine 0.35 (normal = 0.41 ng/ml); urinary norepinephrine 48 mcg/24 h (normal = 66 mcg/24 h), epinephrine 20 mcg/24 h (normal = 24 mcg/24 h); VMA 6 mg/24 h (normal = 1-7 mg/24 h). The investigations of other members of the family were normal.

Discussion

Pheochromocytoma, a chromaffin tumor of the adrenal medulla and sympathetic ganglia is a rare, but curable cause of secondary hypertension in children with an overall incidence of 1% of the cases of childhood hypertension(5). It is particularly dangerous in children because it mimics other clinical conditions(6). However, careful history and radioimaging techniques can help clinch the diagnosis as in the index case. This tumor has been justifiably described as a "pharmacologic bomb", since the sudden secretion of catecholamines from a pheochromocytoma often causes a rapid and dramatic appearance of clinical manifestations(7). Felix Frankel is credited with the first clinical and autopsy report of a pheochromocytoma in 1886(8). In 1927, Charles Mayo was the first to successfully remove an adrenal pheochromocytoma(9). Review of Indian literature reveals that only 6 cases have so far been reported. *Table I* summarizes the clinical features of these children.

Sustained hypertension, recurrent episodes of headache, profuse sweating, weight loss and visual disturbances were the most frequent clinical symptoms and signs in children with pheochromocytoma(6). All three radioimaging studies namely, IVP, ultrasound abdomen and CT scan abdomen in our patient localized the tumor. MIBG (metaiodobenzylguanidine) scintigraphy has a high sensitivity (77%) and specificity (99%). In the literature CT scan has been reported to localize approximately 89% of tumors. Ultrasound delineates up to 80% of lesions and IVP localized the tumor in 60-70% of patients with pheochromocytoma.

Assays of urinary catecholamines and VMA have been associated with approximately 25% incidence of false-negative finding as in our case, whereas such results occur in only 4% of urinary metanephrine determinations(10). Bravo *et al.*(11) reported approximately 35% false-negative results in screening with VMA. The hypertensive crisis in our patient was managed by intravenous infusion of sodium nitroprusside. In one series, 10 of the 14 patients required intravenous sodium nitroprusside for blood pressure control during the acute presentation(12). Use of alpha- and beta-adrenergic blocking agents represents an extremely important addition to the preoperative management. Current blocking regimen consists of phenoxybenzamine, beginning at least one week before surgery. Propranolol

TABLE I–*Review of Children with Pheochromocytoma from India*

Name of author	Age at diagnosis	Mode of presentation	Modality of diagnosis	Outcome
1. Bapna *et al*. 1967(1)	12 years male	Recurrent attacks of severe headache, profuse sweating, convulsions; sustained hypertension	IVP, retroperitoneal pneumogram, Regitin test	Recovered
2. Durairaj *et al*. 1975(2)	12 years male	Dyspnea on effort, palpitation headache, profuse sweating, recurrent vomiting; sustained hypertension	Regitin test, IVP, aortogram	Recovered
3. Durairaj *et al*. 1975(2)	14 years male	Headache, palpitation, sudden hemiplegia, vomiting; sustained hypertension	Regitin test, biochemical tests.	Recovered
4. Durairaj *et al*. 1975(2)	9 years male	Headache, vomiting, dyspnea on effort, palpitation; sustained hypertension	Elevated urinary catecholamine and VMA levels, Regitin test	Died during post-operative period
5. Sharma *et al*. 1987(3)	13 years male	Sustained hypertension	IVP, aortography, biochemical tests	Recovered
6. Sachdev 1990(4)	12 years female	Palpitation, partially calcified thyroid swelling, Diagnosed to have multiple mucosal neuromatosis (MMN) with pheochromocytoma; normotensive	IVP, biochemical tests	Recovered
7. Tomaraei *et al*. 1992 male patient)	9 ½ years	Convulsions, palpitation, headache, abdominal, pain, constipation; sustained hypertension	IVP, ultrasound, CT scan, biochemical tests	Recovered

is added to the regimen three days before surgery(13). After tumor resection, life-long follow-up of urinary catecholamine levels as a screening test for recurrence is recommended. Suspicious results should be evaluated with CT and MIBG scans when available. Careful follow-up allows early detection and treatment of both recurrent and metasatic disease(12).

REFERENCES

1. Bapna BC, Dalla NJS, Singh SM, Bhargava S, Chawla V, Khandpur SC. Pheochromocytoma: An important cause of correctable hypertension. Indian J Pediatr 1967, 34: 410-415.
2. Durairaj M, Sukumar IP, Subramanian N, Krishnaswami S, Mammen KE, Jadhav M. Pheochromocytoma in childhood: A report of 3 cases. Indian Pediatr 1975, 12: 261-265.
3. Sharma BK, Kalra OP, Kataria RN, Khanna SK, Suri S, Chari P, Banerjee AK. Profile of pheochromocytoma in Chandigarh. J Assoc Phys India 1987, 35: 107-111.
4. Sachdev Y. Multiple mucosal neuromatosis. Indian Pediatr 1990, 27: 27-32.
5. Bloom DA, Fonkalsrud EW. Surgical management of pheochromocytoma in children. J Pediatr Surg 1974, 9: 179.
6. Stackpole RH, Melicow MM, Uson AC. Pheochromocytoma in children. Report of 9 cases and review of the first 100 published cases with follow-up studies. J Pediatr 1963, 63: 315-330.
7. Manger WM. Pheochromocytoma. *In:* Conn's Current Therapy. Philadelphia, W.B. Saunders Company, 1991, p 612.
8. Frankel F. A case of bilateral, completely latent adrenal tumor and concurrent nephritis with changes in the circulatory system and retinitis. Arch Pathol Anat Phys 1886, 103: 244-263.
9. Mayo C. Paroxysmal hypertension with tumors of retroperitoneal nerve: Report of a cases. JAMA 1927, 89: 1047-1050.
10. Fonkalsrud EW. Pheochromocytoma. *In:* Pediatric Surgery, 4th edn. Eds Welch KJ. Chicago, Year Book Medical Publishers, 1986, p 1116.
11. Bravo EL, Tarazia RC, Gifford RW, *et al*. Circulatory and urinary catecholamines in pheochromocytoma: Diagnosis and pathophysiologic implications. N Engl J Med 1979, 301: 682-686.
12. Caty MG, Coran AG, Geangen M. Thompson NW. Current diagnosis and treatment of pheochromocytoma in children. Experience with 22 consecutive tumors in 14 patients. Arch Surg 1990, 125: 978-981.
13. Kaufman BH, Telander RL, Van Heerden, J, *et al*. Pheochromocytoma in the pediatric age group: Current status. J Pediatr Surg 1983, 18: 879-884.