A Rare Case of Bilateral Malignant Recurrent Familial Pheochromocytoma

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Abstract

Pheochromocytoma is a adrenal medullary chromaffin cell tumor that typically causes symptoms and signs of episodic catecholamine release, including paroxysmal hypertension. The tumor is an unusual cause of hypertension and accounts for at most 0.1 to 0.2% of cases of high blood pressure. About 90% of pheochromocytomas exist as solitary, unilateral, encapsulated adrenal medullary tumors. About 10% are bilateral, more commonly seen in several members of a family, 40 to 70% of whose members may have bilateral tumors. We report a case of Bilateral Malignant Recurrent Familial Pheochromocytoma .

Case History:

A 21 yr old male patient came to Medicine OPD with H/O: Severe Headache, Sweating, Palpitations & Abdominal Pain since 15 Days. Patient also gave H/o Dyspnea on exertion and mild Chest Pain. Patient is a K/C/O Bilateral Pheochromocytoma since 2000 when he was admitted in GMC Nagpur & Left Adrenal Tumor was removed on 28.02.2000; Patient had Malignant Hypertension with Grade IV Hypertensive Retinopathy. He had recurrence of Pheochromocytoma on Right side which was operated on 24.08.2002 but continued to have Hypertension and his Blood Pressure on admission was 230/140 mmHg and was started on Anti –hypetensive drugs ; Nifedipine 20 mg TDS , Enalapril 5 mg OD & Prazosin 5 mg OD . No Past H/o HTN, DM, PTB, BA, Jaundice. His Younger twin brother died of unknown cause probably Pheochromocytoma as told by relatives and his elder brother also died but cause of death could not be confirmed. On examination Afebrile, Pulse –110 /min, BP – 230/140 mmHg, Pallor + No Icterus / Cyanosis / Clubbing / Lymphadenopathy, JVP – Not raised, NO S/O CCF, .Facio maxillary swelling was seen on right side of the face.

FUNDUS – Grade IV Hypertensive Retinopathy, No E/O any Angiomatous Malformations in the retinal vessels

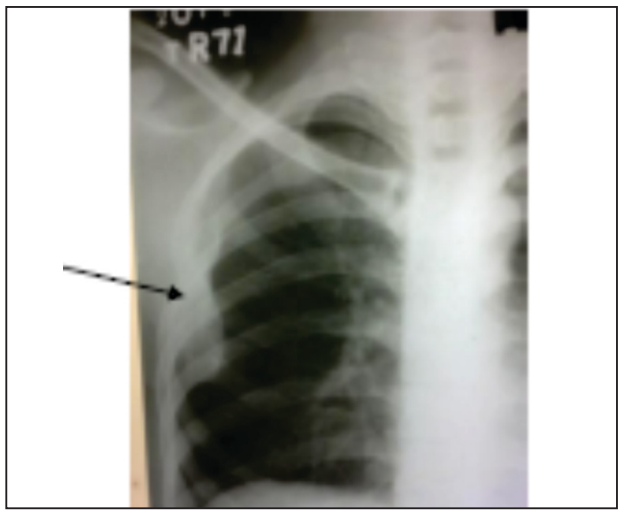
On Systemic Examination Cardiovascular System examination revealed tachycardia, In Heart Sounds – A2 Loud ,No Murmur; Respiratory System :Normal ; Per Abdomen System **Sub costal Scar Marks were seen on both Left & Right Side S/o previously operated**, No Organomegaly / Mass.

CNS: Higher Functions Normal, Conscious oriented, No Focal Neurological Deficit.

Investigations

CBC, KFT, LFT – Within normal limits; Urine Examination: Albumin +++; No RBC / WBC; S.uric acid – 8.9 mg/dl.

ECG – S/O LVH & LAH , **Chest X Ray** – Sclerosis with Expansion



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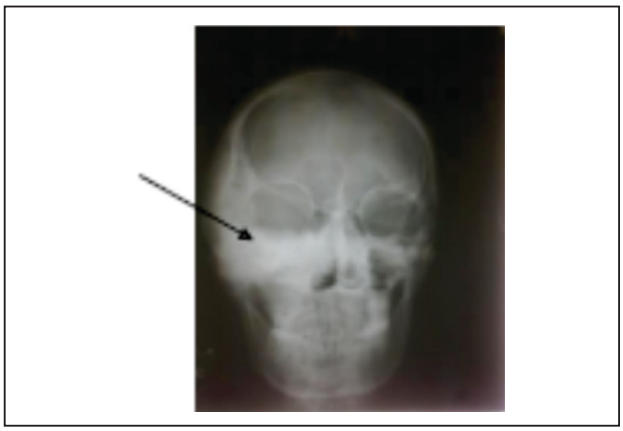
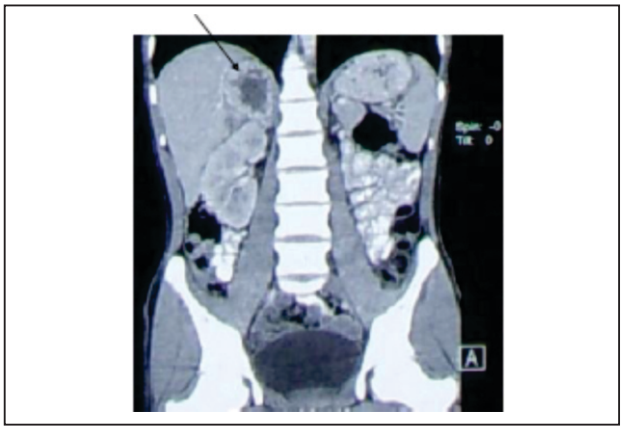
- **X Ray Skull –**Sclerosis in the region of Right Maxillary; Fronto – Zygomatic region with expansion S/o Sclerotic Skeletal Metastasis



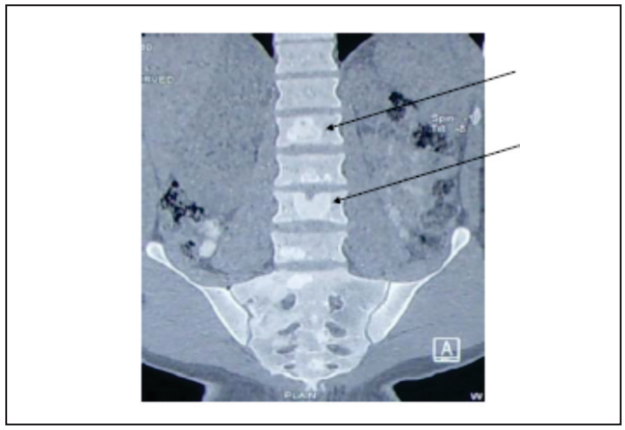
- **CT Abdomen & Pelvis –** A Large Tumor of 7 X 4.2 X 6.2 cm size, well defined, moderately enhancing ,centrally necrotic lesion was noted in the Right Adrenal Region Mildly compressing the superior pole of Right Kidney with multiple lymph nodes in the Pre- Aortic , Para Aortic , Peri Pancreatic and Left Renal Hilar Region.

- There were multiple ill defined sclerotic lesions noted in L2 – L5 vertebral bodies & Right side of sacrum? Bony Metastasis. .



Treatment Given:

- The Oncologist opinion was taken and patient was started on chemotherapy.
- After 6 cycles of chemotherapy, USG & CT ABDOMEN was done &surprisingly the findings remained same.



- Chemotherapy was given with the following drugs: -Palzen ;Vincristine ;Endoxan ; Mesna & Dacarbazine

- Patient is under follow up with us .His Blood pressure is controlled with the above mentioned anti- hypertensive drugs.

Discussion

Pheochromocytoma is a adrenal medullary chromaffin cell tumor that typically causes symptoms and signs of episodic catecholamine release, including paroxysmal hypertension. The tumor is an unusual cause of hypertension and accounts for almost 0.1 to 0.2% of cases of high blood pressure. About 90% of pheochromocytomas exist as solitary, unilateral, encapsulated adrenal medullary tumors. About 10% are bilateral, more commonly seen in several members of a family, 40 to 70% of whose members may have bilateral tumors. Distant metastatic sites include bone, lung, lymph nodes, and liver. Bilateral adrenal medullary hyperplasia has been reported in gene carriers with multiple endocrine neoplasia (MEN) type

2. This hyperplasia may be a precursor of pheochromocytoma. Pheochromocytomas are often described according to the "rule of 10's":

- 10% are extra-adrenal (i.e., 10% would, by common usage, be described as paragangliomas)
- 10% are malignant (15-30% of paragangliomas are malignant)
- 10% are bilateral
- 10% are familial
- 10% are not associated with hypertension

(Although hypertension is a common symptom).

- Other associated factors include multiple endocrine neoplasia (MEN) IIA and IIB, neurofibromatosis type 1, Von Hippel Lindau disease, and familial pheochromocytoma.

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