CASE REPORT

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Retroperitoneal paraganglioma presenting as hypertension and paraparesis in young

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Abstract: Paragangliomas(PGL) are rare tumours that arise from extra-adrenal paraganglia. Extra-adrenal paragangliomas account for only 5 to 10% of all paragangliomas and may present incidentally as a mass. Typical triad of fluctuating hypertension, headache, and sweating is not always present which makes the diagnosis sometimes difficult. Extraadrenal retroperitoneal PGLs are functionally active more often than previously reported and that they are readily detected by soft tissue masses closely associated with the entire length of abdominal aorta. However, no CT feature was found that was unique for PGL.[1] Herein we describe the clinical, radiological and pathological features in young patient presented for hypertension and paraparesis, and later on diagnosed as malignant abdominal paraganglioma with vertebral and lymph node metastasis.

Key words: Paraganglioma; Hypertension; Paraparesis

Introduction

A 15-year-old girl was admitted for paraparesis with bladder involvement since last 3 months' duration. On examination she had significant pallor, hypertension (BP-180/120mm of Hg), left cervical mass 3×3 cm, hard in consistency along with irregular margin and ill-defined masses ranging from 2×2 to 4×4 cm palpable in epigastric region. On CNS examination she had wasting of both lower limbs, power grade 3 with absent knee, ankle and plantar reflexes bilaterally. Rest systemic examination was normal. Her investigations revealed Hb-5gm%, severe microcytic, hypochromic anaemia. Rest biochemical investigations were normal. X ray chest showed superior mediastinal widening. CT (thorax and abdomen) revealed multiple mediastinal and retroperitoneal masses encasing aortic arch, abdominal aorta, IVC and its branches, a 3.8×3.4×2.8cm cervical mass, 4.4×3.4cm mediastinal mass was noted displacing SVC superoanteriorly. Lytic, sclerotic lesions suggestive of metastasis were noted in dorsolumber vertebrae (figure 1), sternum, sacrum, iliac bones and both femoral heads. Remaining intra-abdominal organs were normal. The cervical lymph node was excised and histopathological examination was highly suggestive of metastasis from paraganglioma. Immunohistochemically the tumour cells were strongly positive for Synaptophysin and chromogranin, suggestive of paraganglioma (figure 2). Correlation of these reports with clinical presentation and radiological findings confirmed the diagnosis of malignant paraganglioma. Though urinary catecholamines and genetic studies were strongly indicated in this patient, it was not possible due to financial constraint.

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This patient was started on antihypertensives and other supportive management. Surgery along with radiotherapy was planned but her parents refused to undergo any further management and took her home against medical advice and lost to follow up.

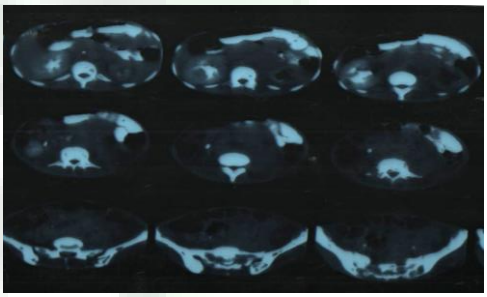

Figure 1: Bony metastasis

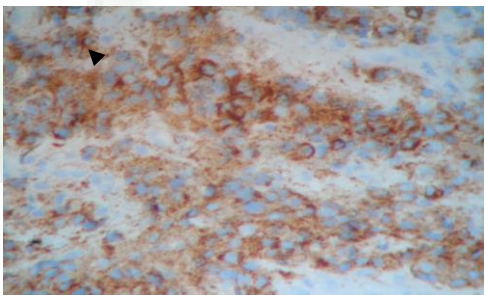

Figure 2: Immunohistochemistry of cervical mass

Discussion

Approximately 5% to 10% of paragangliomas occur in extra-adrenal sites, which can extend from the upper cervical region to the pelvis, parallel to the autonomic nervous system. This distribution

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corresponds to the embryologic development of the paraganglia from neural crest cells.[2] Malignant PGLs defined by the presence of metastasis are very rare and aggressive tumors. The first authors to describe this pathological entity were Miller and Torack in 1970, denominating it a secretary ependymoma,[3] whereas Lerman was the first to coin the term paraganglioma of the cauda equina in 1972.[4] Their study is made difficult by their rarity and the consequent limited number of patients, by their biological variability and variable genetic background. The studies also lack sensitive and specific histopathological and biological markers proving malignancy.[5] The high variability in clinical presentation in PGL is well known.[6] It depends on the variability of these tumours which can express different catecholamines, biosynthetic enzymes, secrete different vasoactive peptides (ANP, neuropeptide Y, adrenomedullin)[7] and present different symptoms related to tumour mass or present different symptoms related to other organ involvement in syndromic forms. Hypertension is the most common feature of secretary PGL (sPGL)). Systemic symptoms like anorexia, fatigue, weight loss and clinical features related to metastatic disease such as pain in bones are common.[5] While histopathological findings are not much useful to differentiate between benign and malignant paragangliomas, extensive local invasion and distant metastasis to liver, bone, and lymph nodes have been used as indicators for malignancy.[5,6,7] sPGLs are usually catecholamine secreting.[8] Malignant PGLs being less differentiated tumors generally secrete noradrenaline and dopamine and has subclinical picture. Metastatic spread may occur at presentation or even many years after primary surgery. Head and neck PGLs are usually clinically silent whereas abdominal sPGLs are usually catecholamine secreting.[9]

Abdominal paragangliomas are mostly retroperitoneal in location, accounting for 85% of all extra adrenal paragangliomas. The most common site for retroperitoneal paragangliomas is between the origin of inferior mesenteric artery and the aortic bifurcation known as organ of Zuckerkandl. Paragangliomas arising from jugulotympanic body are called chemodectomas, whereas paragangliomas originating from the carotid body are known as carotid body tumours. Paragangliomas located in the second part of duodenum are called gangliocytic paraganglioma.[10]

Surgery with complete removal of mass either via laparoscopy or via traditional laparotomy is the treatment of choice for retroperitoneal paragangliomas owning to its malignant potential. Patients with metastatic disease will require adjuvant radiotherapy while chemotherapy is restricted to patients not accessible for surgery and resistant to radionuclide therapy.[11]

References

1. Hayes, W. S. A J Davidson, P M Grimley and D S Hartman Extra adrenal retroperitoneal paraganglioma: clinical, pathologic, and CT findings: AJR American Journal of Roentgenology, 155.6(Dec 1990): 1247-1250.
2. Jaffer Shabnam and Noam Harpaz (2002) Mesenteric Paraganglioma. Archives of Pathology & Laboratory Medicine, 126.3, (March 2002): 362-364.
3. Lerman RI, Kaplan ES, Daman L: Ganglioneuroma paraganglioma of the intradural filum terminale. *Case report J Neurosurg* 36 (1972):652-58.
4. Brodkey JA, Brodkey JS, Watridge CB: Metastatic paraganglioma causing spinal cord compression. *Spine*, 20(1995):367-372.
5. Parenti Gabriele, Benedetta Zampetti, Elen Eapizzi et al: Updated and New Perspectives on diagnosis, prognosis and Therapy of Malignant Pheochromocytoma/Paragangioma. Journal of oncology 2012.10.1155/2012/872713(pubmed)
6. Mannell Mi, L. Ianni, A. Cilotti, A. Conti, "Pheochromocytoma in Italy: a multicentric retrospective study", European Journal of Endocrinology, 141.6(1999): 619-624.
7. Bravo E. L, Evolving concepts in the pathophysiology, diagnosis, and treatment of pheochromocytoma," Endocrine Reviews 15.3(1994): 356-358.
8. Lenders J. W. M., G. Eisenhofer, M., Mannelli & K. Pacak, "Pheochromocytoma." Lancet, 366. 9486(2005): 665-675.
9. Erickson D., Y. C. Kudva *et al.,* "Benign Paraganliomas: clinical presentation and treatment outcomes in 236 patients," Journal of Clinical Endocrinology and Metabolism 86.11(2001): 5210-5216.
10. Disick G. I. S., and M. A. Palese, "Extra-adrenal pheochromocytoma: diagnosis and management," Current Urology Reports, 8.1 (2007): 83–88.
11. Andersen K. F, R. Altaf, A. Krarup-Hansen *et al.,* "Malignant pheochromocytomas and paragangliomas the importance of a multidisciplinary approach," Cancer Treatment Reviews, 37. 2(2011): 111–119.

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