

Case Report

CAROTID BODY PARAGANGLIOMA IN A CHILD: A NOVEL EXPERIENCE

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ABSTRACT

Paragangliomas (PGL) are generally slow growing tumors with unique tissues of origin, location, genetics, potential for biochemical activity, and multi-centricity. Histopathologically, paragangliomas are similar to the adrenal gland neoplasm (phaeochromocytoma). It is usually benign and non-functional. It grows and expands slowly and rarely metastasizes. Since the paraganglionic cells contain very small amounts of catecholamines, clinically significant catecholamine release is rare. Carotid body tumour (CBT) is the most common form of paraganglioma of the head and neck region.

A two and half year female child presented with right sided neck swelling gradually increasing in size in last 6 months and it was present since birth. Radiologically, diagnosis was inconclusive. Histologically, it was diagnosed as paraganglioma which is further supported by immunohistochemistry.

Paraganglioma of carotid body though commonly found in adult but occasionally can be seen in young child. Morphologically, malignant looking tumors should have long term follow up due to metastasis recorded after long interval according to literature. Moreover, in young child hereditary component should be studied if resources available.

Keywords: Carotid body, Paraganglioma, Chromogranin A

INTRODUCTION

Paragangliomas (PGL) are generally benign slow growing tumors with unique tissues of origin, location, genetics, potential for biochemical activity, and multi-centricity. Von Luschka in 1862 and Marchand in 1891 from Europe first described carotid body tumors. Guild in 1953 described vascularized tissue of the convexity of the jugular bulb and promontory of the middle ear which he called glomic tissue. (Myssiorek 2001; Wasserman and Savargaonkar 2001; Baysal 2001)

The nomenclature regarding paragangliomas has been confusing as they have been referred to as glomus tumors, chemodectomas, non-chromaffin tumors, and carotid body tumors. Glenner and Grimely in 1974 clarified this confusion by separating the tumors into adrenal as pheochromocytomas and extra-adrenal tumors as paraganglioma which are then sub classified into carotid and jugulotympanic paraganglioma (both in head and neck region) and viscerautonomic paraganglioma. (Myssiorek 2001; Wasserman and Savargaonkar 2001; Baysal 2001)

Histopathologically, paragangliomas are similar to the adrenal gland neoplasm (phaeochromocytoma); it is usually benign and non-functional. It grows and expands slowly and rarely metastasises (Razakaboay *et al.*, 1999; Casati *et al.*, 1999). Carotid body tumour (CBT) is the most common form of paraganglioma of the head and neck region (Casati *et al.*, 1999). Excess epinephrine or metanephrine should prompt suspicion of an adrenal pheochromocytoma, because head and neck paraganglioma lack the enzyme to convert nor epinephrine to epinephrine (McCaffrey *et al.*).

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A two and half year old girl child presented to the paediatric surgery out-patient department with complaint of swelling in neck. There was no pain or any other complaint associated with it. The swelling is present since birth and now gradually increasing in size in the last 6 months. On clinical examination, the swelling measured 4x4 cm, firm to hard in consistency and non-tender. No palpable enlargement of supraclavicular, cervical or inguinal lymph node was detected. There was no difficulty in swallowing liquid or solid food. She was normotensive. Her total white blood cell count was mildly elevated (13000/mm³). Biochemical investigation for urea, creatinine and random blood sugar was within normal physiological values. Urine examination for vanillyl mandelic acid (VMA) was within normal limits. There was no such relevant family history.

Radiological Findings: CT scan of neck revealed an ovoid, well defined, moderately enhancing space occupying lesion in right lower neck involving the carotid space, pre-vertebral space and extending into superior mediastinum displacing right common carotid artery and internal jugular vein to right and displacing thyroid gland, in trachea, esophagus to left with mild luminal compromise of trachea suggestive of nerve sheath tumour. Ultrasonography reported it as well defined oval, iso to hypoechoic space occupying lesion in anterior triangle of right side of neck, the lesion was showing calcific foci and taking central vascularity branching out peripherally on color doppler study. The lesion was noted to displace carotid vessel and right lobe of thyroid gland.

Operative Procedure: The tumour was successfully removed from carotid vessels under general anaesthesia. There was no major fluctuation in heart rate or blood pressure during intraoperative period. It was loosely attached to artery and could be classified as Group I Shamblin type.

Pathologic Findings: Grossly, tumour was partially encapsulated, deep brown, spongy to rubbery consistency, mostly solid, with occasional areas of cystic change with size of 4.5x3.75 cm.

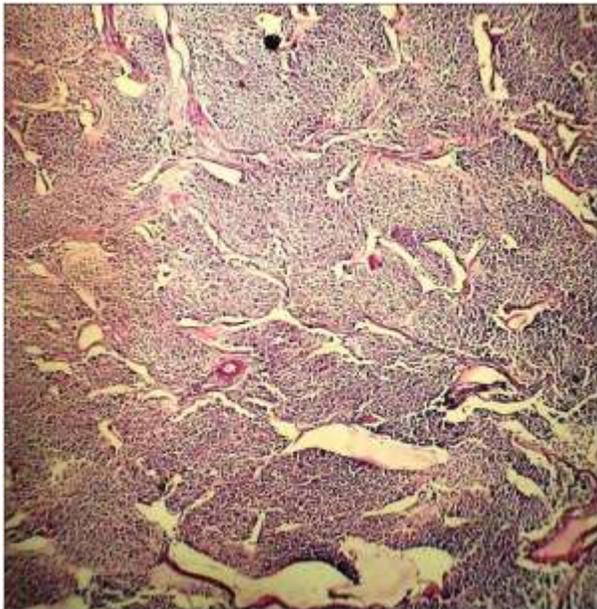


Figure 1: Histopathological examination with routine haematoxylin and eosin stain shows arrangement of cells in well defined nest, classically described as Zellballen pattern surrounded by prominent vascular spaces

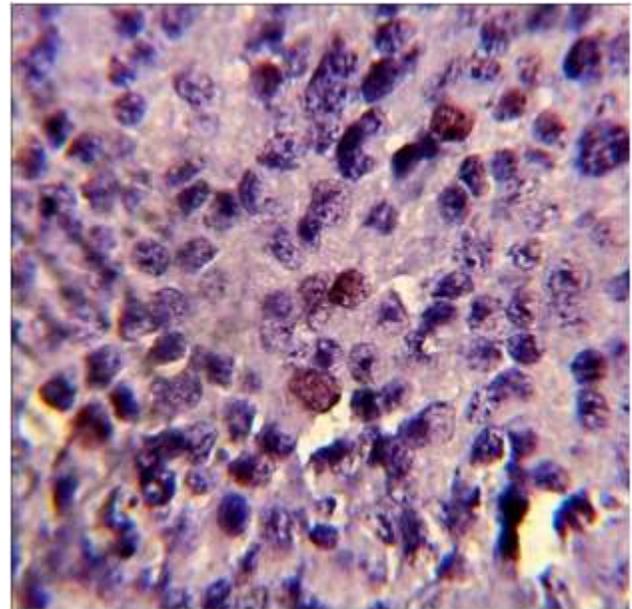


Figure 2: The tumour cells are round to polyhedral, with moderate nuclear pleomorphism, vesicular nuclei, indistinct cell margins with distinct mitosis 16/50 hpf.

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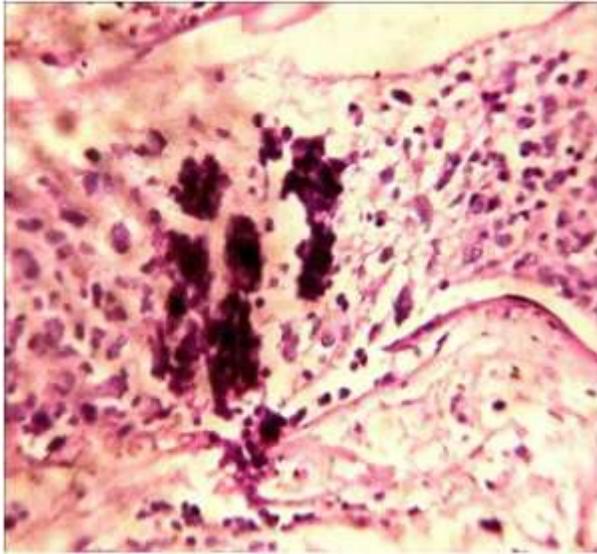


Figure 3: Tumour giant cells are noted in various foci of examination.

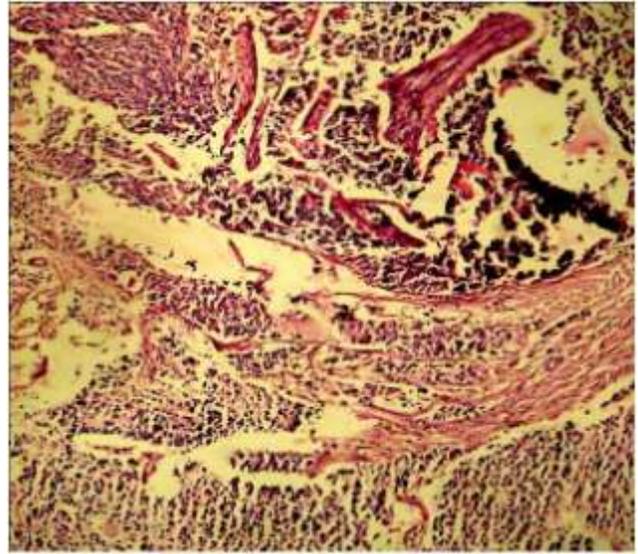


Figure 4: Tumour cells are found to invade blood vessels forming tumour emboli

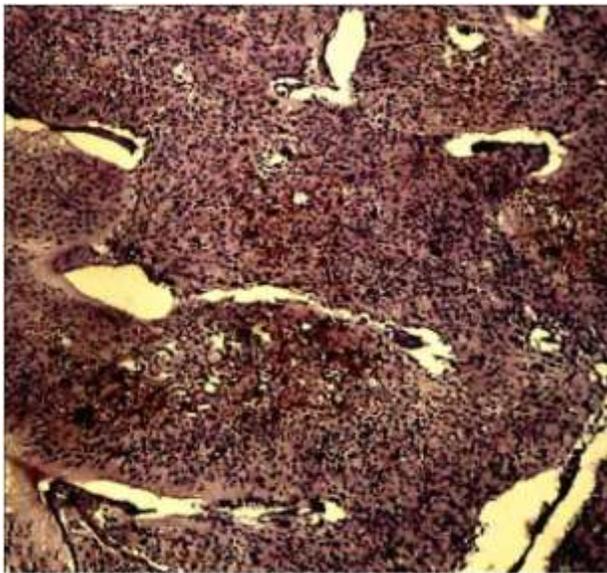


Figure 5: Immunohistochemistry for Chromogranin A shows positive immunoreactivity by tumour cells

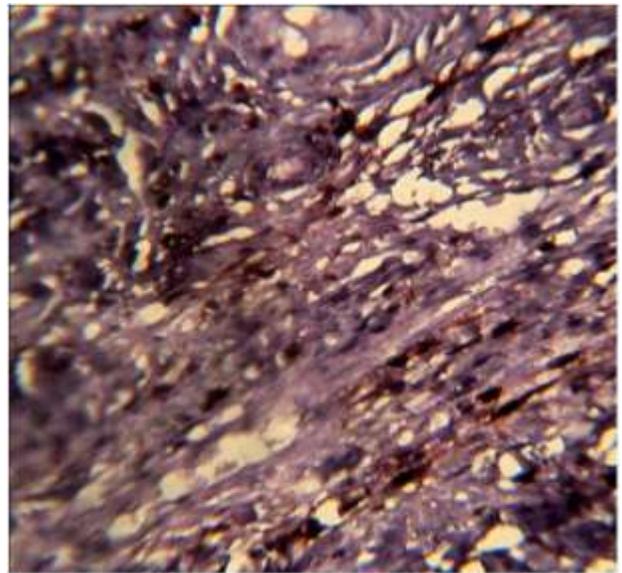


Figure 6: Immunohistochemistry for S100 is positive in sustentacular cells

DISCUSSION

The success in treating benign and malignant PGL is based on early diagnosis, complete resection of the tumour after an adequate catecholamine blockade, which is given prophylactically. Complete resection of the primary mass is the treatment of choice (Barnes *et al.*, 2005). The size of the tumour is important not only for the clinical manifestations, but also for a decision on treatment. In 1971, Shamblin introduced a classification system based on the tumour size, classifying small tumours which could easily be resected from the vessels as Group I. Group II includes tumours that are intimately associated and compressed carotid vessels, but that could be resected with careful subadventitial dissection. Group III consists of tumours that are large and typically encase the carotid artery, requiring complete or partial vessel resection and replacement (Hallett *et al.*, 1998). Between 10% and 50% of PGLs are hereditary

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(autosomal dominant) due to alterations in genes coding for succinate-ubiquinone oxidoreductase subunit D (SDHD), B (SDHB) and C (SDHC) (Grufferman *et al.*, 1980) which map to chromosomes 11, 1 and 1, respectively. Only patients who inherit an SDHD mutation from the father are at risk of developing PGL as the gene causing mutation gets activated during spermatogenesis and not during Oogenesis (genetic imprinting). Parent-of-origin effects are not seen with SDHB or SDHC mutations.

Paragangliomas are rare neoplasms originating from paraganglia within the ganglia of the sympathetic trunk and of the coeliac, renal, suprarenal, aortic and hypogastric plexuses. The paraganglia are clusters of neuroendocrine cells associated with the sympathetic and parasympathetic nervous system. Tumours arising from the chromaffin cells of the adrenal medulla are called pheochromocytomas.

Approximately 1 in 30,000 head and neck tumors is a paraganglioma. They usually occur in the age group of 40-60 years. Malignancy has been reported at all locations of paragangliomas which is determined by metastasis proven through biopsy because paragangliomas may exhibit multicentricity. Local invasion or aggressive behavior do not indicate malignancy of the tumor. (Myssiorek 2001; Wasserman and Savargaonkar 2001; Baysal 2001)

The diagnosis of malignancy reserved for tumours with local, regional and distant metastasis (Davidovic *et al.*, 2005). There are no histologic features that distinguish benign from malignant lesions. This child had the swelling since birth, so chances of having hereditary component are very high. Parasympathetic paraganglia give rise to almost all of the paragangliomas of the upper aerodigestive tract most of which are non-functional, although rare cases exist of adrenocorticotrophic hormone producing nasal paraganglioma associated with Cushing syndrome (Fletcher, 3rd edition).

Irrespective of the site of origin, the histologic appearance of all extra-adrenal paragangliomas is the same. Immunohistochemistry for chromogranin, synaptophysin and S-100 shows reactivity. Rare cases have been cytokeratin-reactive as well (Fletcher, 3rd edition). As the criteria of malignancy does not follow otherwise common criteria like nuclear pleomorphism, mitosis etc. in morphologic ground. But this case showed high degree of pleomorphism with presence of giant cells, frequent mitosis. Some literatures on paraganglioma reported metastasis after 11 long years of resection which requires a follow-up of the present case for the same number of years. After a year, no complaint or recurrence is recorded in the child. It can also be an important case study for gene mutation to establish a hereditary link to tumorigenesis.

Conclusion

PGL of carotid body though commonly found in adult but occasionally can be seen in young child. Morphologically malignant looking tumors should have long term follow up due to metastasis recorded after long interval. Moreover, in young child hereditary component should be studied if resources available.

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