Paraganglioma of Right Cervical Plexus—A Case Report

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Abstract Head and neck paragangliomas (HNPs) and pheochromocytomas are rare tumors. Catecholamine secreting tumors that arise from chromaffin cells of the adrenal medulla and the sympathetic ganglia are referred to as pheochromocytomas and extra adrenal catecholamine secreting paragangliomas (extra adrenal pheochromocytomas) respectively. The distinction between pheochromocytoma and paraganglioma is an important one because of implications for associated neoplasms, risk for malignancy, and genetic testing. Paragangliomas are rare tumors of the head and neck and are of diagnostic challenge. We report a case of 55 year old male who had a neck swelling, cough with expectoration, which on extensive evaluation revealed to be Paraganglioma with primary optic atrophy.

Keywords Paraganglioma · Cervical plexus · Optic atrophy · Zellballen pattern · Pheochromocytoma

Case History

A male patient aged 55 years was admitted to the medical ward for the treatment of bronchitis 2 months ago when a swelling was noticed on the right side of his neck which was asymptomatic in nature.

On Examination

A 3 × 4 cm swelling was seen on the right side of the neck at the level of lower 1/3rd of SCM. It was immobile, firm in

consistency. Swelling was compressible, increased in size on valsalva maneuver (Figs. 1, 2) and decreased in size on contraction of the SCM. Transmitted pulsations felt. There was no local rise of temperature or tenderness.

Investigations

The following investigations were done:

Routine investigations
Liver function tests
Thyroid profile
VMA

The above were within normal limits

Ophthalmology examination revealed bilateral primary optic atrophy.

CT Scan

Brain: Normal

Neck and Thorax: (Fig. 3) A significantly enhancing soft tissue mass seen adjacent to the right lobe of the thyroid at the level of the bifurcation of the brachiocephalic trunk deep to the SCM. Right Para tracheal and supraclavicular lymphadenopathy noted. Features were suggestive of paraganglioma.

USG Neck

A well defined lobulated highly vascular mass seen in the right lower neck with:

- Tortuous vessels along the periphery
- Splaying of the proximal CCA and Vertebral artery

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Fig. 1 Swelling before valsalva maneuver

Fig. 2 Swelling after valsalva maneuver

- The mass is causing lateral displacement of the thyroid and proximal CCA
- No cervical lymphadenopathy
- A well defined nodule in the left lobe of thyroid.

Treatment

Excision Under GA

- Incision
- Elevation of skin flaps
- Division of the Sternocleidomastoid muscle
- External Jugular vein and Common Carotid artery identified

Fig. 3 Arrows showing significantly enhancing soft tissue mass

Fig. 4 Arrow shows a spindle shaped mass seen arising from a nerve trunk lateral to the Common Carotid Artery and just superior to the Brachiocephalic trunk

- A spindle shaped mass seen arising from a nerve trunk lateral to the CCA and just superior to the Brachiocephalic trunk (Fig. 4).

Fig. 5 Excision of the mass

Fig. 6 Dissection carried out from medial to lateral

Fig. 7 Field after excision

Fig. 8 Cut specimen with nerve trunk

- Excision (Figs. 5, 6, 7).
- Pleura of the right lung was intact.
- Vagus nerve palpated deep to Common Carotid artery.
- Specimen (Fig. 8) sent for HPE.

Post Operative Period

Post operative period was uneventful

There were no signs of carotid sinus syndrome.

2 pints of cross matched blood was transfused.

Patient was mobilized on the first post operative day.

HPE

HPE showed epithelioid cells arranged in Zellballen pattern with a (Fig. 9) fibrous surrounding layer of sustentacular cells with interspersed blood vessels.

Discussion

Approximately 10 % of Head and neck paragangliomas and pheochromocytomas have been cited to be malignant. Benign as well as malignant HNPs consist of the same two major cell types: epithelioid chief cells and sustentacular supporting cells. These two cell types form clusters of

Fig. 9 Histopathology showing fibrous surrounding layer of sustentacular cells with interspersed blood vessels

cells, so-called *Zellballen*, that are surrounded by extensive vascular sinusoids. Interestingly, no accepted pathological or immunohistochemical markers distinguish malignant from benign paraganglial tumors [1]. The distinction between pheochromocytoma and paraganglioma is an important one because of implications for associated neoplasms, risk for malignancy and genetic testing. Paragangliomas are found where there is chromaffin tissue, along the Para aortic sympathetic chain, within the organs of zuckerkanal (at the origin of the inferior mesenteric artery), in the wall of urinary bladder and along the sympathetic chain in the neck or mediastinum. Odd locations for paragangliomas include the intra-atrial cardiac septum, spermatic cord, vagina, scrotum and sacrococcygeal region. Paragangliomas in the head and neck region (carotid body tumors, glomus tumors, chemodectomas) usually arise from parasympathetic tissue and typically do not hyper secrete catecholamine and metanephrines. Whereas paragangliomas in the mediastinum, abdomen and pelvis usually arise from sympathetic chromaffin tissue and usually do hyper secrete catecholamine and metanephrines. Familial paragangliomas are an autosomal dominant disorder characterized by paragangliomas that are located most often in the head and neck but also in the thorax, abdomen, pelvis and urinary bladder. Approximately 5 % of head and neck paragangliomas and more than 50 % of abdominal paragangliomas produce hormones. Most cases of familial paragangliomas are caused by mutations in the succinate dehydrogenase (SDH; SDHB, SDHC, SDHD). The prevalence of familial paragangliomas is unknown. Druck et al. [2] had found 19 % incidence of malignancy. Regional lymph nodes and lungs are common sites of secondaries.

Paraganglioma syndromes may be increased in those who live in high altitudes or are chronically exposed to hypoxic conditions. Avoidance of habitation at high

altitudes and activities that promote long-term exposure to hypoxia should be considered. Activities such as cigarette smoking that predispose to chronic lung disease should be discouraged in persons who have a mutation in *SDHD*, *SDHC* or *SDHB*. By age ten years or at least ten years before the earliest age at diagnosis in the family, pre-symptomatic testing, including genetic testing, should be offered to all first-degree relatives of an individual in whom a mutation in *SDHD*, *SDHC* or *SDHB* has been detected. Use of molecular genetic testing for early identification of at-risk family members improves diagnostic certainty and reduces the need for costly screening procedures in those at-risk family members who have not inherited a disease-causing mutation [3].

Conclusion

Approximately 30 % of apparently sporadic HNPs are caused by a germ line mutation in one of the *SDHB*, *SDHC* and *SDHD* genes. We therefore recommend molecular genetic screening of all HNP patients for those mutations. First-degree relatives of mutation carriers have to be screened as well. There is an autosomal-dominant role of transmission for all PGLs with a parent-of-origin-dependent inheritance in *SDHD* mutation carriers (PGL 1). Patients with PGL 1 and 4 have a very high lifetime risk for the development of HNP as well as thoracic and abdominal paraganglionic tumors. Compared with sporadic HNP, tumors in *SDHB*, *SDHC* and *SDHD* mutation carriers develop at a significantly younger age. The *SDHB* mutations are associated with a high percentage of malignant PGLs. Three-body regions clinical screening of mutation carriers offers the chance to diagnose those tumors in an early and asymptomatic stage. Appropriately timed surgical intervention will minimize disease-specific morbidity and mortality.

We present a case of Head and neck paraganglioma with primary optic atrophy where early detection through surveillance and removal of tumors may prevent or minimize complications related to mass effect hyper secretion, and malignant transformation.

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