

Non-functional cervical paraganglioma in an adult: A case report

Pradeep Balineni¹, Sandeep Pathivada¹, Samuel Dev Merlin¹, Shruthi Kamal¹

From ¹Saveetha Medical College and Hospital, High Road, Chennai, India

Correspondence to: Dr. Pradeep Balineni, Saveetha Medical College and Hospital, High Road, Chennai - 600077, Chennai, India.

E-mail: pradeep052191@gmail.com.

Received - 26 February 2019

Initial Review - 13 March 2019

Accepted - 09 April 2019

ABSTRACT

Paragangliomas are rare neuroendocrine tumors. The most common site is the adrenals cervical region. Cervical paragangliomas are divided into carotid body and vagus types. Here, we report the case of a non-functional cervical paraganglioma in a 78-year-old male came with complaints of swelling in the left side of the neck for 2 years. On imaging, guided core needle biopsy showed eosinophilic cells with vacuolated nuclei and some inclusion bodies which suggested a possible clear cell tumor, neuroendocrine tumor or a paraganglioma. Complete surgical excision was done under general anesthesia, postoperative histopathology and immunohistochemistry markers (S100, Synaptophysin, and Ki67) suggested of paraganglioma.

Keywords: *Non-functional, Paraganglioma, Surgical excision, Vagal.*

Paragangliomas are rare, vascular, neuroendocrine tumor arise from the neural crest cells. The annual incidence of paragangliomas is 3-8 cases per 1 million population per year, which comprises 0.6% of all head and neck tumors^{1,2}. They may be functional (catecholamine secreting) or non-functional. Functional tumors show an abundance of extra-adrenal chromaffin stained cells. The most common site for extra-adrenal paragangliomas is head and neck. Head and neck paragangliomas are divided into vagal nerve (Xth nerve) and carotid body types. Surgery of these tumors remain the mainstay of treatment, radiotherapy can be tried as adjuvant therapy [1,3]. We present the case of a non-functional cervical paraganglioma in a 78-year-old male. We are reporting this case in view of its rarity in the incidence.

CASE REPORT

A 78-year-old male presented with complaints of swelling in the left side of the neck for 2 years. Swelling rapidly increased in size in the past 6 months. There were no other complaints of pain fever, any other swellings. There was no history for any neural deficit, hypertension, diabetes mellitus, and tuberculosis. There were no bowel and bladder disturbances.

On examination, patient general condition and vitals were found to be stable. On local examination of the neck, a 14x10 cms swelling was noticed in the posterior triangle (Fig. 1). The swelling was smooth surfaced, hard in consistency and skin over the swelling appeared normal.

The computed tomography (CT) imaging revealed a hypodense lesion with hypervascularity in the posterior triangle abetting the sternocleidomastoid muscle and the trapezius muscle with

no infiltration into any vital structures (Fig. 2). An ultrasound-guided core needle biopsy was done which showed eosinophilic cells with vacuolated nuclei and some inclusion bodies which suggested a possible clear cell tumor, neuroendocrine tumor or a paraganglioma.

Under general anesthesia, the patient was taken up for complete surgical excision of the tumor (Fig. 3). Postoperatively, the patient did not have any complaints of swallowing difficulties or any neural deficit. On histopathological examination and immunohistochemistry marker study of the postoperative specimen, it was seen positive for S100, Synaptophysin, and Ki67 suggesting a paraganglioma (Fig. 4). On a one month follow-up, the patient was comfortable and asymptomatic.

DISCUSSION

Kohn coined the term 'paraganglioma' in the year 1900. They may be inherited or sporadic [3]. In the case of familial paragangliomas, they are inherited in an autosomal dominant manner. They can be seen along with hereditary syndromes such as PGL1, PGL3, and PGL4 which occur due to mutations in SDHD, SDHC, SDHB genes respectively, whereas PGL2 syndrome has no specific mutation [3,4]. Usually, they are benign in 85-95% of the cases [4]. Till now, 93 cases of paragangliomas were reported to have a malignant transformation in the USA [1] as these tumors are generally locally aggressive with rare distant metastasis. An estimated mean growth time for paraganglioma is 1.0mm/year and a mean doubling time of 4.2 years [2,4]. WorldHealth Organization (WHO) classified them as intra-adrenal and extra-adrenal tumors. In extra-adrenal head and neck are the common sites. They constitute to about 0.6% of all head and neck tumors.

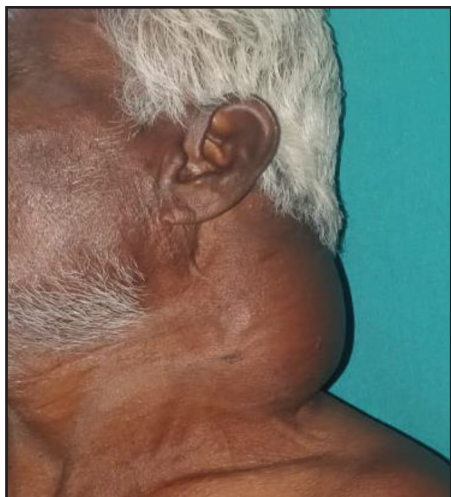


Figure 1: Swelling in the lateral aspect of the neck

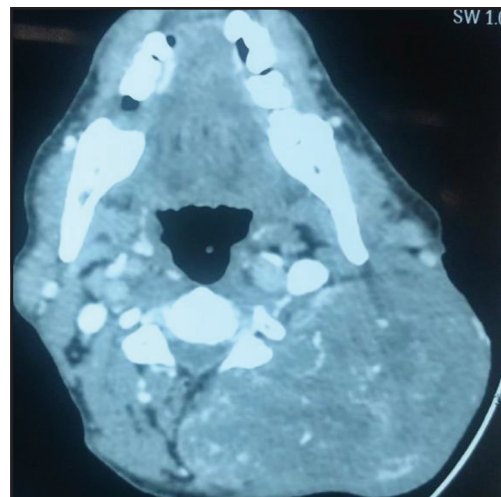


Figure 2: CT image showing the swelling

Head and neck paragangliomas are broadly divided into vagal and carotid body types. Netterville classification and Shambin classification can be used for further classifying vagal and carotid body tumors respectively [4]. Netterville classification classifies them as type A (tumor confined to neck), type B (tumor in contact with jugular foramen) and type C (tumor penetrating into or extending beyond jugular foramen with or without intracranial extension). Shamblin classification classifies these tumors as type I (not involving carotid artery), type II (partially involving internal carotid artery or carotid body), type IIIA (completely involving internal carotid artery or carotid body with no skull base contact) and type IIIB (completely involving internal carotid artery or carotid body with contact to skull base) [4]. The differential diagnosis for this condition is pheochromocytoma, atypical carcinoid of the larynx, alveolar soft part sarcoma, melanoma, glomus tumors.

Paragangliomas are generally solitary and present in the 5th decade of life. The youngest case reported was of 5 years age [5]. They are found to be multicentric in 30-40% of familial cases and 10% of sporadic cases [1]. Clinically, paragangliomas are divided into functional and non-functional tumors, 4% of tumors are also reported to be hyperfunctional [6]. Functional tumors present with

features of hypertension, headache, palpitation, fever, pallor, dyspnea, vomiting, dizziness, flushing, seizures and paraesthesias, whereas, non-functional tumors are generally found incidentally or may present as slow-growing tumors [3]. Approximately, 30% of vagal tumors and 2% of carotid body tumors may present with nerve deficit also [4].

On ultrasonogram, they are seen as heterogeneous, hypoechoic, hypervascular lesions and on contrast-enhanced computed tomography (CECT), they appeared as hypodense lesions with rapid, intense contrast enhancement and early venous washout. Carotid body type tumors show a characteristic lyre appearance on CT [4]. Magnetic resonance imaging (MRI) would be the standard imaging modality for paragangliomas, in which they are seen as hypo or isointense lesions on T1 weighted images and hyperintense on T2 weighted images [7,8]. MRI angiography can be done to evaluate for vascular displacement [8]. Core needle biopsy would show two types of cells: type 1 shows catecholamine granules and type 2 cells similar to Schwann cells [1]. On immunohistochemistry, staining these tumors stain positive for CD56, synaptophysin, chromogranin, NSE, S100 and negative for Keratin, EMA, mucin, CEA, TTF1, HMB45 [9].

The various treatment modalities used for paragangliomas are surgery, radiotherapy, wait and watch methods, with surgery

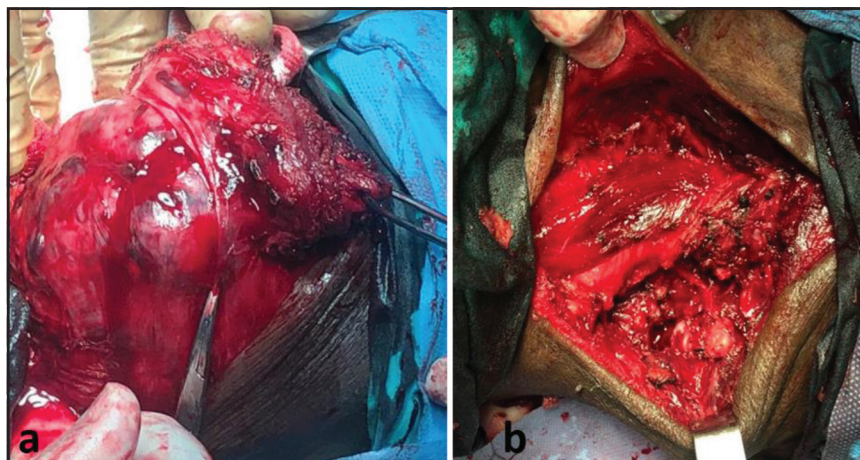


Figure 3: (a) The intraoperative picture; (b) bed of the tumor

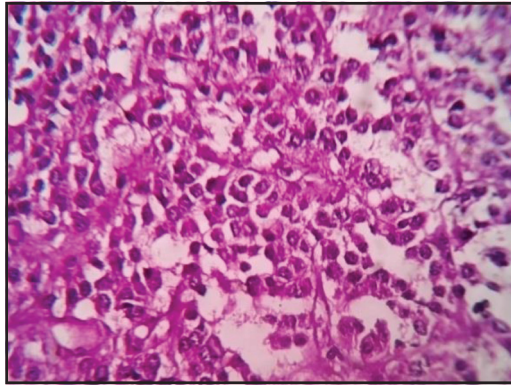


Figure 4: The histopathological slide with eosinophilic cells and vacuolated nuclei

being the standard treatment for these. But, each case should be individualized before taking up for any treatment [2,3,7]. Complete surgical excision of the tumor would give a cure rate of 89-100% [2]. In case of functional tumors, alpha and beta blockers are given to counter with hypertension prior to surgery [3]. A preoperative embolization can also be tried 2-3 days prior to surgery to reduce the intraoperative bleeding [4]. Lymphadenectomy is generally not done but in case of lymph node metastasis, regional lymphadenectomy can be done. Approximately, 60-80% of cases have swallowing difficulties in the immediate postoperative period, but only 14% of cases have a permanent postoperative morbidity [4] and 30% of the cases have a recurrence even after complete surgical excision [3]. Gamma knife and Cyberknife radiotherapy are given in cases of large, non-resectable tumors as an alternative to reduce the tumor load and to make the tumor resectable [2,4]. Henerman *et al.* quoted 96-100% tumor control by radiotherapy with a dose of 45Gy². There is no possible evidence in support of postoperative radiotherapy. Tumors which are nearby large vessels or important structures and are non-resectable can be waited and watched as the mortality rate in non-treated cases with age more than 70 years is less than 10% [4]. Some authors also suggest palliative chemotherapy

by cyclophosphamide, dacarbazine, and vincristine in cases of non-resectable tumors [3].

CONCLUSION

Cervical paragangliomas are very rare tumors and difficult to diagnose. Surgical excision is the treatment of choice.

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Funding: None; Conflict of Interest: None Stated.

How to cite this article: Balineni P, Pathivada1 S, Merlin SD, Kamal S. Non-functional cervical paraganglioma in an adult: A case report. *Indian J Case Reports*. 2019;5(2):171-173.

Doi: 10.32677/IJCR.2019.v05.i02.026