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Hypopharyngeal paraganglioma: a case report

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Summary
Paragangliomas are neuroendocrine neoplasms that primarily afflict patients during the fourth and fifth decades of life. The majority of extra-adrenal paragangliomas arise in the head and neck region, notably from the carotid and aortic bodies, jugular bulb and tympanic plexus. Although one-fifth of all parapharyngeal neoplasms are paragangliomas, primary hypopharyngeal paraganglioma is relatively uncommon, only one other case having previously been documented by Filippin *et al.* (1989). We report a second case of hypopharyngeal paraganglioma in an 18-year-old male as seen in our center.

Keywords: *Paraganglioma, hypopharynx, tracheostomy, Ibadan, Nigeria.*

Résumé
Les paragangliomes sont les néoplasmes neuro-endocrine qui primairement afflictent les patients Durant les 4ieme et 5ieme années de vie. La majorité des paragangliomes extra-adrenal commence par la tete et le cou notamment de la carotide et de l aorte, de la bulbe jugulaire et le plexus tympanique. Bienque un-cinquiéme de tous les néoplasmes paraphyrangéales sont les paragangliomes, le paraganglíome hypopharayngeal primaire n'est pas relativement commun sauf le seul cas documenté par Filippin et al (1989) . Nous avons reporté un second cas de paragangliome hypophyyngéal chez un male de 18 ans dans notre centre.

Introduction

Paragangliomas are neuroendocrine neoplasms derived from autonomic paraganglia [1,2]. The head and neck is the principal site for extra-adrenal paragangliomas. Most head and neck paragangliomas arise from the carotid and aortic bodies, jugular bulb, and tympanic plexus, and are distinctively much less frequently encountered in the upper aerodigestive tract [1,2,3]. However, neoplasms of the pharynx are relatively uncommon, and paragangliomas constitute an overall 20% of all parapharyngeal neoplasms [3].

A search of the medical literature reveals that Filippin *et al* 1989 have previously reported a case of hypopharyngeal paraganglioma [4]. It is the purpose of this paper to record a second case of hypopharyngeal paraganglioma.

Case report

An eighteen-year-old male presented at the accident and emergency unit of University College Hospital, Ibadan with a 20-month history of muffled voice, foreign body sensation in the throat and in respiratory distress. There was a history of progressive dysphagia, which became most severe in the few days preceding his presentation.

He was dehydrated and emaciated. Throat examination revealed a huge, immobile mass, almost completely shutting off the view of the hypopharynx on indirect laryngoscopy. A lateral soft tissue x-ray of the neck revealed a huge broad based globular soft tissue shadow in the hypopharynx extending from C_2 to C_5 (Fig. 1).

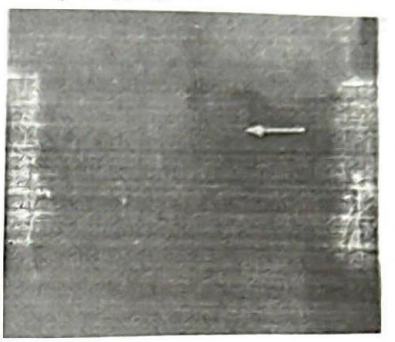

Fig. 1: Lateral soft tissue x-ray of the neck revealing a huge broad based globular soft tissue shadow in the hypopharynx extending from C_2 to C_5

He had tracheostomy under local anaesthesia, thereby securing the airway and ensuring a safe anaesthetic induction. Examination under anaesthesia (EUA) showed a firm, well-encapsulated mass attached by a broad stalk to the right posterolateral wall of the hypopharynx. A wedge biopsy of the lesion resulted in a torrential bleed, which was safely controlled.

Histological examination of the resected specimen revealed a highly vascularised cellular neoplasm composed of sinusoidal clusters of uniform cuboidal to polygonal cells with abundant granular cytoplasm (Figures 2 and 3).

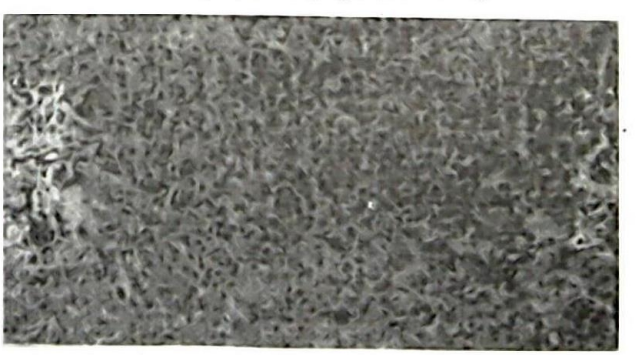

Fig. 2: Histological section of the neoplasm revealing sinusoidal clusters of uniform cuboidal to polygonal cells with abundant granular cytoplasm (Haematoxylin and cosin, X40)

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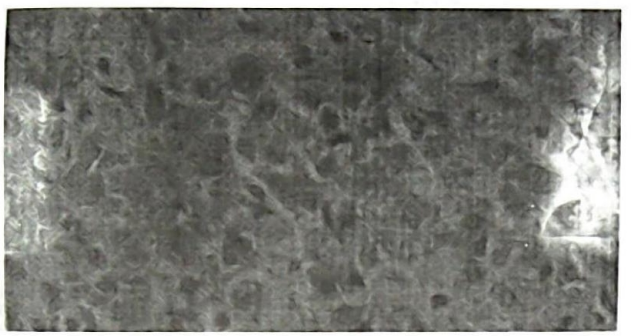

Fig. 3: Higher magnification of neoplasm emphasising morphological details of the tumour cells (Haematoxylin and eosin, X70)

Immunostaining revealed granular diffuse cytoplasmic positivity for neuron specific enolase, features consistent with a paraganglioma. The 24-hour urinary vanillylmandelic acid level (VMA) was assayed following the histological diagnosis. The level was normal (1.3mg/L; 2.0mg/24 hrs.).

Subsequently, through a horizontal collar incision and infrahyoid right lateral pharyngotomy approach, the lesion was completely excised, histology of which confirmed the original diagnosis. He was discharged 14 days after having been decannulated 7 days post operatively. The patient has remained free of any recurrence five years after.

Discussion

Paragangliomas arise from paraganglionic cells (paraganglia) of neural crest origin, which migrate in close association with the autonomic ganglion cells. Embryologically, the paraganglia arise from neural crest and migrate symmetrically on either side of the midline, to extend from the middle ear region and the base of the skull to the pelvic floor. The paraganglionic system is divided into the adrenal medulla and the extra-adrenal system. Anatomically, paraganglia are further divided into three; namely the branchiometric, intravagal and aorticosympathetic groups. The trachea, larynx, and hypopharynx belong to the aorticosympathetic group. They are innervated by the parasympathetic nervous system, and therefore they rarely secrete catecholamines. Because they are responsive to oxygen and carbon dioxide tension within adjacent tissues, they are sometimes called chemodectomas [2,3,5]..

The review of Ferlito *et al* [6] demonstrates that paragangliomas are uncommon, presenting mainly during the fourth to sixth decades of life. They are three to four times more common in females, and show no racial predilection. The rarity of paragangliomas in general and of laryngeal and tracheal paragangliomas, in particular, makes this case of hypopharyngeal paraganglioma worth recording.

The clinical features associated with paraganglioma may be vague and long standing; requiring a high index of suspicion coupled with relevant investigations to arrive at a diagnosis. An initial lateral soft tissue x-ray of the neck is valuable. This may show soft tissue shadow obstructing the airway as it was in this case. However, computerized tomography and magnetic resonance imaging are the best radiological means of determining the nature and extent of paraganglioma [2].

The clinical differential diagnoses of hypopharyngeal paraganglioma include atypical carcinoid tumour, malignant melanoma, medullary and anaplastic carcinoma, neurofibroma and chondrolipoma. These other entities are easily distinguished from paraganglioma on morphological grounds. Macroscopically, paragangliomas are encapsulated, tan-grey to purple-red tumours, which may be haemorrhagic or spongy. Histologically, they are composed of chief cells and sustentacular cells with a peripheral fibrovascular stromal layer that are organised into a whorled pattern referred to as "zellballen" [3,7].

As demonstrated in our index case, severe bleeding is the most common complication of histological diagnosis and therapy [7]. Thus, adequate preparation should be made in this regard. Surgical excision is the mainstay of treatment for paraganglioma [7,8,9]. An infrahyoid right lateral pharyngotomy approach was used in this case as it ensured adequate resection of the tumour and control of bleeding. The patient has been followed up for five years now without any evidence of recurrence.

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