Parapharyngeal Ganglioneuroma of Hypoglossal Nerve in a 4 Year Old Girl: a Rare Case Report

Neelakamal Hallur · B. C. Sikkerimath ·
Santhosh S. Gudi · Girish Chour · Sneha Ginimav ·
Madhumati Nidoni

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Abstract Parapharyngeal ganglioneuroma of hypoglossal nerve is very rare benign tumor arising from sympathetic nervous system producing mass and functional effect. We present a rare case in 4 year old girl with history of swelling and odynophagia in left side of neck. Extensive clinical, immunohistochemistry, and imaging of the swelling confirmed the diagnosis of ganglioneuroma after surgical excision.

Keywords Parapharyngeal space · Ganglioneuroma · Hypoglossal nerve

Introduction

Ganglioneuromas are fully differentiated benign, neurogenic tumors arising from central or peripheral components of the sympathetic nervous system. They are quite rare; with the location of the cervical region for this tumor has a reported incidence of only 1–5% in the literature [1]. Majority are diagnosed in patients older than 10 years of age in the posterior mediastinum followed by retroperitoneum and adrenal proper. Ganglioneuromas are slow growing, noninvasive lesions that are often detected incidentally as a painless mass or because of a mass effect on neighbouring structures. Ganglioneuromas arising in the pharynx and neck are uncommon. Cervical sympathetic ganglioneuroma is one of the rarest of neck tumors [2]. We present a parapharyngeal ganglioneuroma of hypoglossal

nerve in a 4 year old girl and discuss its diagnosis and management.

Case Report

A 4 year old girl child was referred to department of oral and maxillofacial surgery with 1½ year history of a gradually increasing painless swelling on the left side of neck and 1 year history of difficulty in swallowing and foreign body sensation in the throat. There was no history of hearing loss or dyspnea. General physical examination did not reveal any pathological findings and vitals were within normal limits. Examination of the swelling revealed a soft non tender mass measuring 7 × 4 × 3 cm extending from left lower border of mandible, left ear lobule and supraclavicular region superoinferiorly and from midline of neck to posterior border of sternocleidomastoid mediolaterally. Skin over the swelling was normal and no local rise in temperature was noticed. Preoperatively there was mild deviation of the tongue on the left side making us suspicious of hypoglossal nerve involvement by the tumor. Rest of the cranial nerve functions was normal with no signs of Horner's syndrome, or any other neurologic deficits (Fig. 1a). Routine lab tests were within normal limits. Chest X-ray was normal. Tests for urine vanillylmandelic acid were negative thus ruling out adrenal gland involvement. There was no history of hypertension, diarrhea, sweating or flushing associated with the mass ruling out sympathetic involvement. Fine Needle Aspiration Cytology (FNAC) report showed individually scattered and small clusters of cuboidal cells with round nuclei and moderately abundant eosinophilic cytoplasm suggestive of carotid body tumor (paraganglioma). Ultrasonography (USG) with 7.5 MHZ linear probe showed a well defined

N. Hallur (✉) · B. C. Sikkerimath · S. S. Gudi · G. Chour ·
S. Ginimav · M. Nidoni
Department of Oral and Maxillofacial Surgery, PMNM Dental
College and Hospital, Bagalkot, Karnataka, India
e-mail: neelakamallhallur@hotmail.com; omfsbgk@gmail.com

Fig. 1 **a** Preoperative clinical view of the patient note: a well defined swelling on the left side of the neck. *Inset* shows extensions of the swelling from dorsal view. **b** Postoperative clinical view showing the persistence of tongue deviation on left side suggestive of hypoglossal nerve involvement by the tumour

Fig. 2 (**a**, **b** and **c**) Preoperative CT of neck axial and coronal view showing well defined lobulated soft tissue density mass in the carotid sheath of left parapharyngeal space

capsulated hypo echoic rounded mass lesion along left carotid sheath with multiple fine calcifications within the lesion. Doppler investigation of the mass showed increased vascularity, giving an impression of enlarged lymph node or neurogenic tumour due to calcifications in the mass. Further investigation with computed tomography of neck (plain and contrast) revealed a well defined, lobulated, heterogeneously enhancing soft tissue density mass lesion arising from the left carotid space, situated in left parapharyngeal space, measuring $3.3 \times 3.7 \times 5.4$ cm. Multiple non enhancing hypo densities were also noted in the mass. Craniocaudally, the lesion extended from the skull base up to left supraclavicular fossa. Anteriorly the lesion was related to left CCA, ICA, ECA, submandibular gland, medial pterygoid muscle, and prestyloid parapharyngeal space fat thus ruling out the origin of the mass from parotid gland. Medially the mass was abutting the pharynx with mass effect over it leading to odynophagia. Posteriorly, the mass was related to the left erector spinae muscles, cervical vertebrae and left IJV. Laterally the mass was abutting left

strap muscles and deep lobe of left parotid gland. Left CCA, ICA, ECA, was anteriorly displaced by the mass and left IJV was being displaced posteriorly. Interestingly right jugular vein was dilated measuring 17 mm probably because of compression of left IJV due to the mass (Fig. 2a–c). These findings left us with the impression of schwannoma or paraganglioma. We also suspected neurogenic origin of the tumour since it is in this region that neurogenic tumours are common. MRI revealed preservation of soft tissue planes surrounding the tumor suggesting non invasive lesion.

CT angiography of neck showed a not well demarcated, non-homogeneously enhancing soft tissue density mass lesion in the left carotid space involving predominantly pre-styloid parapharyngeal space and partly extending into posterior cervical space, it measured $4.8 \times 3.8 \times 3.7$ cm and extended 2 cm below skull base to supraclavicular region in craniocaudal direction. Mass was posterior to carotid vessels displacing and compressing the left IJV postero laterally. It did not show any evidence of

Fig. 3 **a** Preoperative CT angiography of neck showing anteriorly displaced carotid vessels and posterolaterally compressed left internal jugular vein and feeder vessel being the ascending pharyngeal artery. **b** Surgical exposure of the mass and its relation with vital structures of the neck. *Inset* shows resected specimen

calcification, but focal areas of necrosis were noted. No evidence of invasion was seen. Mass was supplied by branches of left ECA mainly ascending pharyngeal artery, with no significant abnormality in carotid and vertebral artery (Fig. 3a).

Surgical excision of the tumor was done by a transcervical approach. Subplatysmal flap was raised, external jugular vein ligated. Sternocleidomastoid was dissected out bluntly and was retracted to reveal carotid sheath with its contents IJV, ICA, CCA, and ECA. ICA, CCA, and ECA were splayed medially and IJV was displaced laterally by the tumor. Carotid sheath was bluntly dissected to expose the tumor between IJV and CCA. With careful sharp dissection, the tumor was dissected from its contents in anterior, posterior, and medial margins of the lesion. Hypoglossal nerve was found stretched at the superior border of the tumor confirming the origin of this neurogenic tumor. The mass was freed carefully from its attachment to the hypoglossal nerve at the base of the skull and was taken out in toto. Other structures identified were spinal accessory nerve, vagus nerve, superior thyroid artery, lingual artery, and facial artery which were then preserved in continuity (Fig. 3b). As expected tongue deviation to left side was noticed due to involvement of hypoglossal nerve. After 1 month of follow up deviation of the tongue still persists (Fig. 1b).

Histopathology

Gross pathology revealed well encapsulated, smooth mass, which was firm in consistency, oval in shape and pink-tan in color and measured $6 \times 4 \times 2.3$ cm in its dimensions. Microscopically, in the H. & E. stain the lesion was encapsulated, well circumscribed, and homogeneously cellular. The lesional cells were elongated or spindle shaped with wavy nuclei (Schwann cell-like), were arranged in the form of streaming fascicles and intersecting

Fig. 4 **a** 200 $\times$: Showing S-100 positive spindle and oval tumor cells. *Inset* shows weakly positive ganglion cells for chromogranin

arrangements. Interspersed within these cells were scattered and few clusters of large round to oval cells with abundant coarse eosinophilic cytoplasm and nuclei ranging from 1 to 3 suggestive of ganglion cells were also seen. Immunohistochemical evaluation confirmed S-100 positivity of the lesion and weak positivity of ganglion cells for chromogranin (Fig. 4).

Discussion

GNs are derived from primordial neural crest cell that migrate from mantle layer of developing spinal cord and populate primordia of sympathetic ganglia and adrenal medulla [3].

GNs are rare benign neural origin tumors when compared with schwannoma and neurofibromas. They were

first described by Leretz in 1870 and was reported first as occurring in neck by Dequervain in 1899 [4]. GNs occur generally in older children with slight female predominance of 1.5:1 with mean age of presentation being 7 years and are typically slow growing [5]. Cervical sympathetic chain is the most common site of origin. Other sites of origin are postmediastinum, retroperitoneum, larynx, pharynx, nodose ganglion of vagus nerve and very rarely the parapharyngeal space, hypoglossal nerve, intervertebral foramen, and spinal canal [2, 4].

Parapharyngeal GNs in pediatric population are exceedingly rare [4] and to the best of our knowledge only one case of hypoglossal nerve ganglioneuroma has been reported till date. Being non invasive, the location of sympathetic chain in the neck affords the tumours of neck an opportunity to readily spread into parapharyngeal or retropharyngeal space. Symptoms could be because of compression of vital structures by the enlarging mass or when it is a functioning tumor, e.g., compression of pharyngeal wall, cranial nerve, eustachian tube, or vascular structures. Patient may complain of dyspnea, dysphagia and foreign body sensation, and snoring or obstructive sleep apnea leading to misdiagnosis of tonsillitis or peritonsillar abscess. Eustachian tube obstruction can be suspected if there is serous otitis or conductive hearing loss with palpable neck mass. Pulsations over the swelling can be as a result of intrinsic tumor vascularity or transmission from internal carotid artery. In our case there were false pulsations visible and palpable medial to the mass. Symptoms due to functional tumors can be hypertension, diarrhea, and sweating, flushing and renal acidosis. Diarrhea has been related to presence of vasoactive intestinal peptides found in cytoplasm of mature ganglion cells. Some patients show elevated levels of urine vanillylmandelic acid. Patients may also exhibit Horner's syndrome if origin of tumor is within cervical sympathetic chain [4, 6].

GNs can occur in association with Turner's syndrome, Horner's syndrome, multiple endocrine neoplasia carcinoma, pheochromocytoma, GI disorders, Marfanoid facies or multiple ganglioneuromas [4, 6–8].

A judicious combination of USG, FNAC, CT, MRI, and angio CT delineates the location, nature, and extent of tumor and facilitates treatment planning [5].

Once diagnosis of GN is confirmed surgical excision alone is curative. In case of large mass where complete excision is not possible debulking of the mass can be planned. Various surgical approaches to this space are transcervical–transparotid, transcervical, orbitozygomatic middle fossa, and transmandibular approach and combinations. Transcervical is considered to be an optimal

approach for neural origin tumours and minor salivary gland tumors [9]. Recurrence rate of GNs is generally considered as negligible. GNs rarely undergo malignant transformation to malignant schwannoma or malignant peripheral nerve sheath tumor.

Parapharyngeal ganglioneuromas are rare benign tumors of neurogenic origin, usually noted as enlarging parapharyngeal or cervical masses in the oropharynx or neck. Characteristic pathology and location in prestyloid region confirms the benign tumor. Accurate clinical workup with investigations helps in correct diagnosis and treatment planning. It is possible to treat parapharyngeal ganglioneuromas, with minimum intra-op vascular and post-op neural complications by utilizing transcervical approach.

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