

Case Report

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A 3-year-old child presenting with a cervical mass: diagnostic dilemma

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ABSTRACT

Ganglioneuromas are benign tumors that originate from the sympathetic chain and are composed of ganglion cells and nerve fibres. These are most often located in the mediastinum (20%), retroperitoneum (10%), and adrenal gland (30%). We report a case of a 3-year-old female child presenting with a neck mass and ptosis for 4 months. A computed tomography (CT) scan of the neck was done, which showed well defined ovoid lesion in the neck suggestive of a neurogenic tumor. Image-guided fine-needle aspiration cytology (FNAC) was done, which was given as a ganglioneuroma. With imaging and histopathological guidance, we have done meticulous excision under general anaesthesia (GA). Although ganglioneuromas are common neurogenic tumors, their occurrence in the cervical region is very rare. Ganglioneuromas should be considered as part of the differential diagnosis for pediatric soft tissue tumors of the head and neck. The diagnosis for ganglioneuromas in the cervical region can only be ascertained with postoperative pathologic examination, and excision is considered the only effective treatment modality known so far, which may cause Horner's syndrome at times. However, patients have a favorable prognosis without recurrence overall.

Keywords: Ganglioneuromas, Neck mass, Paediatric tumors, Surgical management, Neurogenic tumors, Horner's syndrome

INTRODUCTION

Ganglioneuromas are benign differentiated tumors of neural crest derived cells in the autonomic nerves. A ganglioneuroma is a fully differentiated, benign counterpart of a neuroblastoma.¹ They are rare, affecting one per million of the population. Most are sporadic, but they can be associated with neurofibromatosis type II and MEN 2B. Ganglioneuromas are most frequently found in the posterior mediastinum and retroperitoneum with the involvement of the cervical region being very uncommon. When present in the neck, they may manifest as a swelling, pain, or symptoms related to compression of the upper aerodigestive tract.² They are composed of an admixture of ganglion cells and Schwann/stroma cells. They develop in childhood but typically present later as they are non-secreting and slow growing.

Its diagnosis often requires surgical biopsy, since fine needle aspiration is often inconclusive and imaging

findings of ganglioneuromas are variable and nonspecific.³ Excision is the treatment of choice. The prognosis is excellent as recurrences are rare.

We here report a case of 3-year-old presenting with cervical mass and ptosis with no significant past medical history. This study aimed to include ganglioneuroma as a differential diagnosis for paediatric cervical masses. Also the need for its meticulous excision for excellent prognosis.

CASE REPORT

A 3-year-old child's primary complaint when they arrived at our outpatient department (OPD) was a 4-month-old lump in the lateral portion of their right neck. Pain, a history of dysphagia or dyspnea, or a change in voice were not linked to mass. For a month, the patient experienced ptosis. No noteworthy medical history in the past. Vital signs and a general check-up were normal. Upon local

examination, there was a single, firm, non-tender lump measuring 4×2 cm in the right upper and lateral neck region; the skin above it was normal.

Standard laboratory tests were conducted and are within acceptable bounds. A well-defined ovoid lesion on the right side of the neck, anterolateral to the carotid at the level of bifurcation, was seen on a contrast-enhanced computed tomography (CT) scan of the neck. It had a well-defined capsule, was heterogeneous in enhancement, and did not infiltrate surrounding structures. It is causing the right internal jugular vein to shift laterally. This lesion was suggestive of a neurogenic tumor, which arises from the nerves within the carotid sheath. A biopsy with image guidance suggested ganglioneuroma.



Figure 1: Axial CT image of neck with contrast showing a right latero cervical mass (yellow arrow) with heterogeneous enhancement and central hyper enhancement in relation to carotids and jugular vein (red arrow) and trachea (green arrow) at the level of T1.

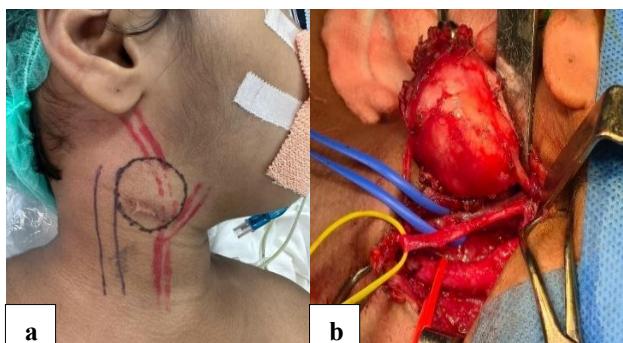


Figure 2: (a) Preop skin marking of the tumor, and (b) intraop dissection of tumor from surrounding vital structures.

The lump was completely surgically removed using a transcervical technique while under general anesthesia. Under the sternocleidomastoid muscle in the right lateral neck, a solitary, oval, well-circumscribed tumor measuring 5×3 cm was discovered by physical examination. The internal jugular vein was somewhat displaced laterally, and the lesion was situated directly over the carotid sheath. The edges of the mass were carefully dissected away from

the neck's contents using blunt and sharp instruments. The procedure went well, and there were no problems throughout the recovery phase.

Upon gross examination, a single soft tissue mass measuring $4.5 \times 2.7 \times 2.6$ cm with a greyish white cut surface was found to be well-circumscribed and capsulated. A microscopic analysis revealed a mixture of mostly Schwannian stroma and sporadic nodules of mature ganglion cells. The Schwann cells exhibit moderate nuclear atypia; mitosis is inconspicuous. No definite immature neuroblasts noted. Pathological diagnosis of ganglioneuroma was made.

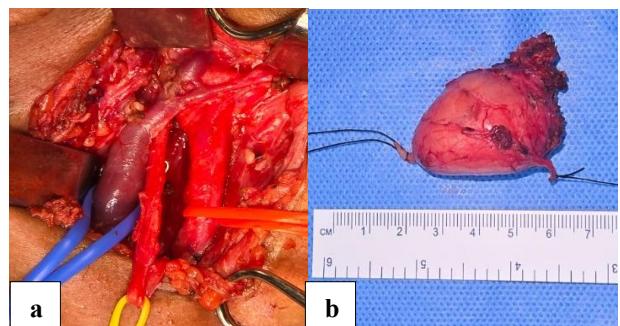


Figure 3: (a) Visualisation of vital structures, after tumor excision, and (b) macroscopic appearance of the tumor.

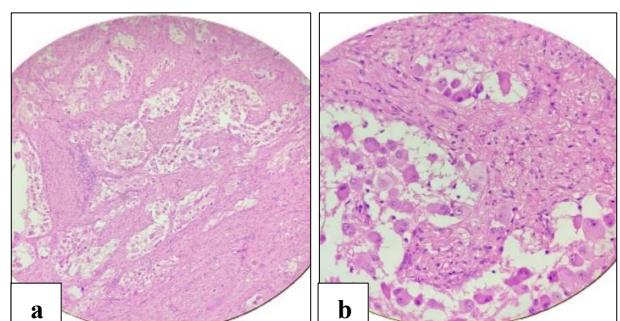


Figure 4: (a) Low-power view (H&E, $\times 20$) showing a well-circumscribed tumor composed of mature ganglion cells and spindle cells (H&E, $\times 20$), and (b) high-power view (H&E, $\times 40$) demonstrating mature ganglion cells with eccentric nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm, interspersed with spindle-shaped Schwann cells.

DISCUSSION

Most neurogenic tumors arise from the cells of the nerve sheath, from ganglion cells, or from the paraganglionic system. The incidence, cell types, and risk of malignancy strongly correlate with patient age. Ganglion cell tumors (ganglioneuromas, ganglioneuroblastomas, and neuroblastomas) arise from the sympathetic chain or from the adrenal medulla.

Ganglioneuromas are well differentiated benign tumors characterised histologically by well differentiated ganglion cells with a background of schwann cells, these are most often found incidentally in asymptomatic young adults. GN are typically sited in the thoracic cavity mainly posterior mediastinum (60–80%), the abdominal cavity (10–15%), adrenal gland, retroperitoneum, pelvic, sacral and coccygeal sympathetic ganglia, and the organ of Zuckerkandl, and the cervical region (5%).^{4,5} Other less common locations are the middle ear, the parapharynx, the skin, the orbital space, and the gastrointestinal tract.^{6,7}

Cervical ganglioneuroma is an uncommon, benign neurogenic tumor, accounting for approximately 6% of pediatric tumors.⁸ This condition was first described by Loretz in 1870.⁹ Most ganglioneuromas present as asymptomatic masses in the cervical region. However, symptoms may occur due to compression of nearby anatomical structures. Common clinical signs include difficulty swallowing, shortness of breath, changes in voice, and neck pain. Other possible symptoms include features of Horner's syndrome, such as ptosis, miosis, anhidrosis, and facial flushing, which typically result from injury to the cervical sympathetic chain. In some cases, systemic effects like hypertension, excessive sweating, diarrhea, and metabolic disturbances such as renal acidosis may develop due to catecholamine secretion, which can increase levels of vanillylmandelic acid (VMA) or homovanillic acid (HVA).¹⁰

Once a mass is discovered CT and MRI are usually performed to characterise the lesion. They are well defined with a capsule and calcification may be present. GNs have a low T1 and a high T2 weighted signal on MRI. Radiology cannot definitely diagnose a GN; histology is required and so preoperative diagnosis can be challenging.

Complete surgical excision is the treatment of choice.¹¹ The excision goals are to confirm the diagnosis, to stop the tumor growth and subsequently relieve the compression of anatomic structures. GN are usually of good prognosis. The risks are related to preoperative injuries of adjacent neural and vascular structures. Follow up imaging is mandatory, although recurrence after total excision is rare.¹²

CONCLUSION

The prognosis for ganglioneuromas, which are benign, well-differentiated neuroblastic tumors, is usually favorable. Despite their rarity, they ought to be taken into account when making a differential diagnosis for cervical masses. It can be difficult to make a firm diagnosis prior to pathological evaluation because there aren't any clear clinical symptoms or distinguishable imaging

characteristics. Therefore, for an accurate diagnosis and suitable treatment, knowledge of ganglioneuromas and their appearance is crucial.

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