



PHARYNGEAL GANGLIONEUROMA: A RARE CASE REPORT

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ABSTRACT

Ganglioneuromas, benign tumors of the sympathetic nervous system, are rare to arise in the parapharyngeal space. They are usually asymptomatic and hormonally silent. The interest of this case lay in the rarity of its incidence at the pharyngeal space. Majority of cases are detected incidentally during work-up for unrelated conditions. They usually develop from the great sympathetic chains, extending from the base of the skull to the neck, posterior mediastinum, retroperitoneum, and adrenal glands.

We report, case of a 48 years old male and discuss, imaging feature and the histopathological data.

Keywords: Ganglioneuroma, Ganglion cell, Pharynx, Computed Tomography.

INTRODUCTION

Ganglioneuroma is a rare and benign tumor of the autonomic nerve fibers arising from neural crest sympathogonia, which are completely undifferentiated cells of the sympathetic nervous system. However, ganglioneuromas themselves are fully differentiated neuronal tumors that do not contain immature elements. [1]

Ganglioneuroma can occur anywhere, posterior mediastinum accounted for about 41.5%, retroperitoneum 37.5%, adrenal gland 21%. Few GNs occur in the cervical region (8%), of which the majority are in single form. [2] It also occurs at other parts like spermatic cord, heart, bone and gastrointestinal tract.

Currently, histopathologic examination is the only tool to diagnose ganglioneuroma and to differentiate it from other neural crest tumors.

Report of a case:

The patient, a 48-year-old male, presented at our hospital with a complaint of weakness, weight loss and intermittent fever for three months. ENT examination showed slightly congested left pharynx, no swelling of the epiglottis and vocal cords. The patient has no neurologic deficit. The laboratory studies were unremarkable. For detail study and doubtful symptom patient was advised for CT scan which revealed a focal tumor with soft tissue density, 26 x 32 x 45 mm in size and well defined border is shown in the parapharyngeal space in CT scan. Plain CT shows uniform density (isodense with the surrounding muscle, fig 1), clear, oval mass. On enhanced CT scan the tumor showed obvious enhancement, and showed higher density than the density of surrounding muscles (fig 2). It shows linear cloud like enhancement in small parts of tumor. No tumor necrosis is identified.

The top of the tumor is in the jugular foramen, and the appearance of the tumor was gray with slight fibrous covering. The interior is strongly adherent to the internal and external carotid arteries, and the internal carotid artery is compressed. The internal jugular vein is pushed outward and 2cm below the jugular foramen. The position of the tumor is close to the internal jugular vein and the vagus nerve travels through the tumor. The inner hypoglossal nerve is at the parenchymal border of the tumor.

Ganglioneuromas are circumscribed and they appear to be encapsulated. Histologically, they consist of ganglion cells, axonal processes in large numbers, satellite cells and Schwann sheaths. Myelination is absent. The tumor was diagnosed as ganglioneuroma with well differentiated ganglion cells and neural sheath cells. It also contained fibrous component and abundant mucous matrix which originated from the sympathetic nerve. No mitotic activity, necrosis, or calcification was present.

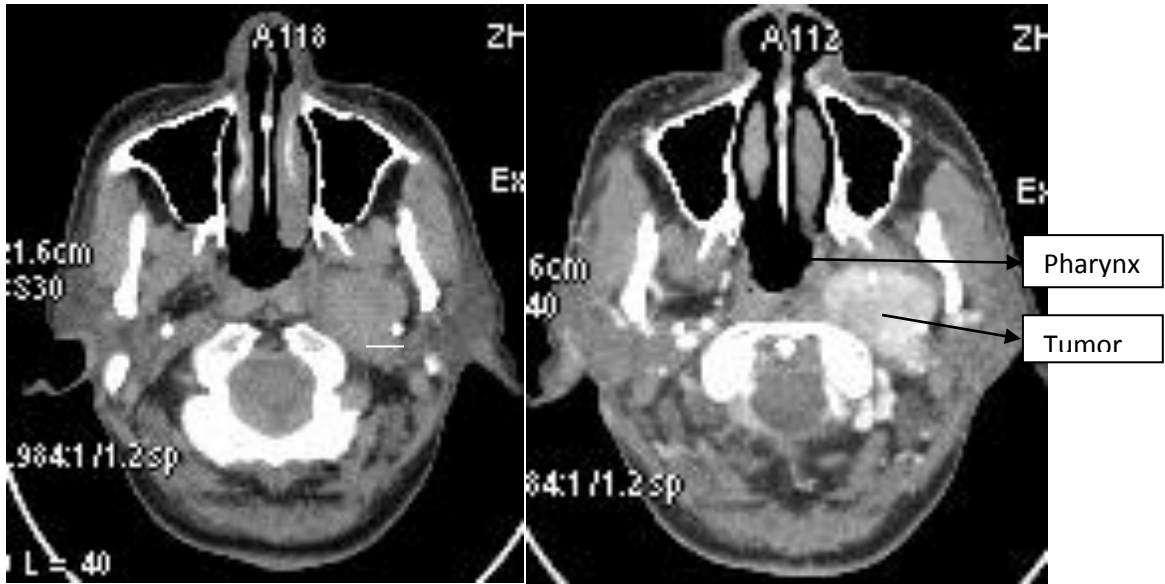
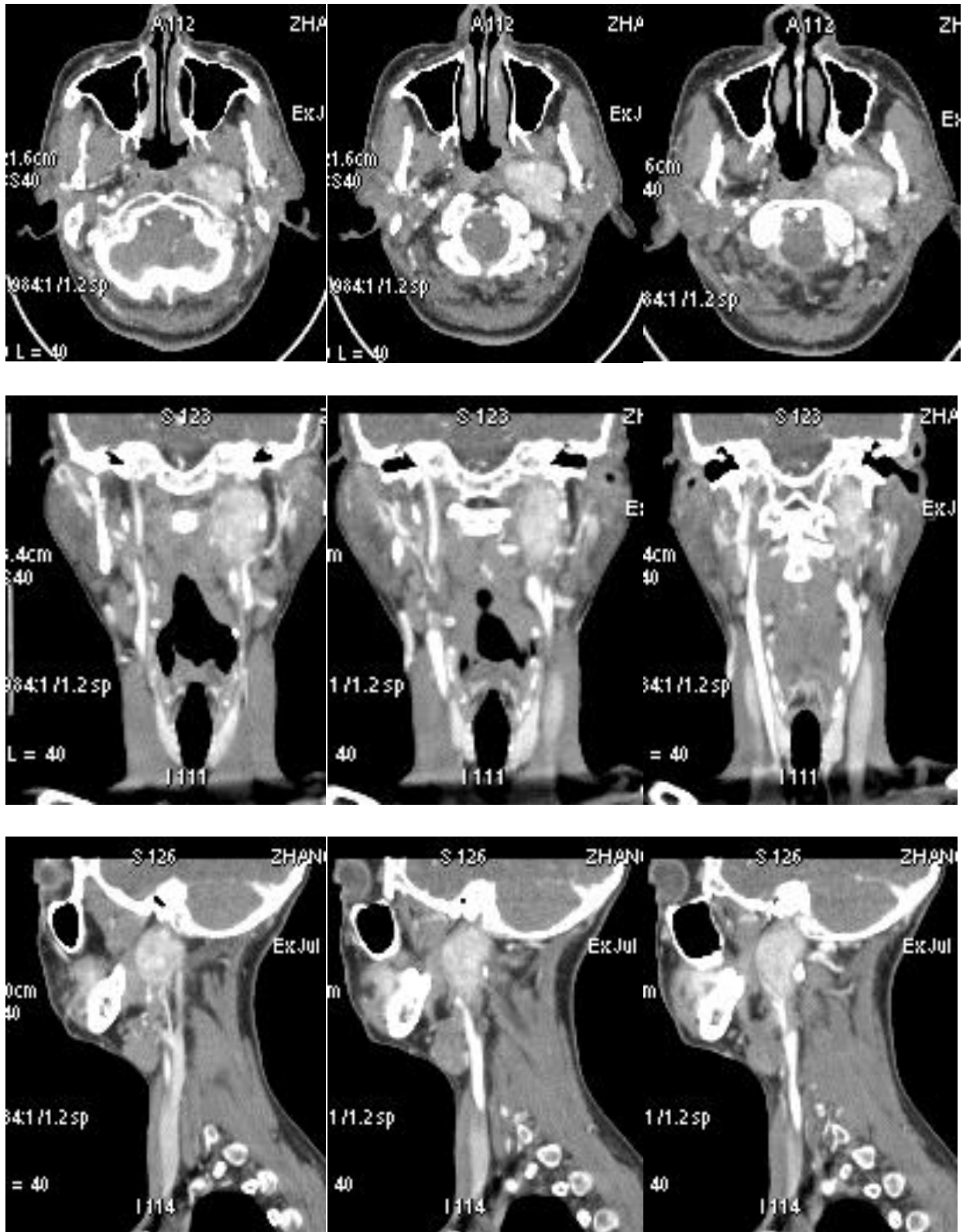


Figure 1: Plain CT showing isodense tumor with surrounding muscles.

Figure 2: Enhanced CT showing hyperdense tumor.



Enhanced CT images showing pharyngeal ganglioneuroma.

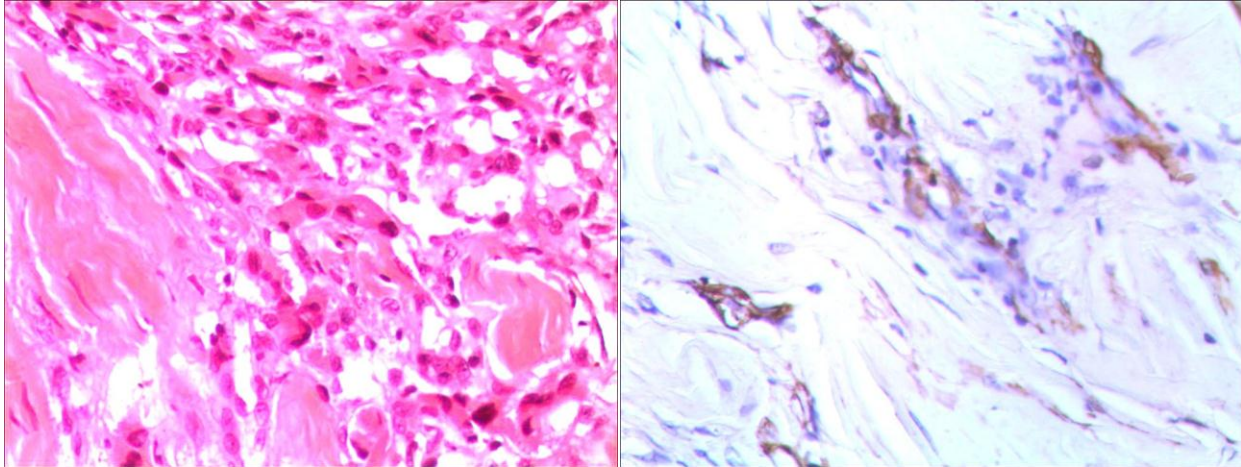


Figure 3: Histopathology of ganglioneuroma showing ganglion cells, Schwann cells and fibrous tissue.

DISCUSSION

Ganglioneuroma is a rare, benign, non-invasive and neurogenic tumor. It is a well-differentiated tumor of the sympathoadrenal nervous system. These tumors belong to a family of neoplasms that exhibit a wide range of differentiation, with neuroblastoma at one end and ganglioneuroma at the other. GN in the head and neck region usually present as a slowly enlarging mass, predominantly in single form. Only one case of the multiple form in one side of the neck has been reported; thus, it is extremely rare. [2] The most frequent sites of presentation include the posterior mediastinum and the retroperitoneum. In general, the neck is a rare site of presentation. [3] We describe a case of ganglioneuroma observed in an unusual location, the parapharyngeal space.

Ganglioneuroma can be found anywhere along the sympathetic chain, but its most common localizations are the posterior mediastinum and retroperitoneum. They occur over a wide age range (0-70 years), with the adrenal tumors tending to occur in patients older than 10 years. Also occurs on other parts like spermatic cord, heart, bone and gastro-intestinal tract. The clinical behavior of ganglioneuroma is invariably benign, although the occurrence of chronic diarrhea due to tumor elaboration of vasoactive intestinal peptide has been described. [4] Moreover, catecholamine synthesis is frequently observed in this tumor, which rarely induces hypertension as a clinical sequela.

The hypotheses for the pathogenesis of benign ganglioneuromas include the spontaneously or artificially induced maturation of neuroblasts in a neuroblastoma into distinct ganglion cells, separation of the remaining cells from the embryonic neural crest and necrosis of neuroblasts at an early stage of tumor

development. [5]

The development of ganglioneuroma is characteristically silent, and the appearance of clinical symptoms depends on the tumor size and site. These tumors most often manifest as an asymptomatic mass originating from the cervical sympathetic chain.

Ganglioneuromas are considered to have a low level of secretory activity or no secretory activity, but even in the case of activity, patients are rarely symptomatic. [6]

The most characteristic histologic feature of ganglioneuromas is the presence of mature ganglion cells. Macroscopically, ganglioneuromas appear to be pale, encapsulated, although a true capsule is infrequent. Microscopically, the tumors are composed of intersecting bundles of spindle cells resembling neurofibromas or neurilemmomas, loose myxoid stroma and dysplastic ganglion cells that resemble normal ganglion cells both morphologically and immunohistochemically. Immunohistochemistry can be helpful in ascertaining the origin of the tumor and the differential diagnosis, but it is not essential for diagnosis.

Malignant transformations of ganglioneuromas are exceptionally rare, possibly because growth regulation factors derived from Schwann cells have a protective influence on malignant tumor cells. [7]

In this context, the various other possible differential diagnoses for a cervical mass are carotid body tumor, neck hemangioma, cervical lymphoma, cervical lymph node tuberculosis, lymph node metastasis, schwannoma, neuroblastoma, lipoma.

Surgical excision is the treatment of choice. Numerous approaches for resection of neoplasms in the parapharyngeal space have been described, including the transoral, transcervical, transparotid, transcervical-transpharyngeal, and the infratemporal fossa approach and combinations of these. The location, size and pathological type determine the choice of the surgical approach. Our patient underwent transcervical excision.

Differential Diagnosis:

- A. Carotid body tumor:** It is located in the common carotid artery bifurcation so that the external and internal carotid artery fork angle increases also called as Lyre sign. It has soft tissue density similar to muscle. It also shows significant bright and rapid enhancement in contrast enhanced CT.
- B. Jugular venous tumor:** It occurs in the skull base jugular vein and its vicinity. Plain scan is of soft tissue density which gets significantly enhanced in contrast scanning.
- C. Neck hemangioma:** It is found in subcutaneous or deep neck usually in infants and young children, often capillary hemangioma.
- D. Cervical lymphoma:** It is one of the common malignancy in children that arise from mature

lymphocytes. Single or bilateral multiple enlarged lymphnodes. The mass is usually larger.

- E. Cervical lymph node tuberculosis:** Often multiple, matted type lymph nodes, most frequently seen in age group of 11 to 30. Affected nodes show central caseation. Also shows ring enhancement due to central necrosis surrounded by inflammatory region or may appear frankly cystic.
- F. Metastasis:** Metastatic tumors are very common in the late stages of cancer. Lung, breast, skin, colon, liver, cervix are common site of origin. History, sign and symptoms reveals no any significant near and distant primary tumor.
- G. Schwannoma:** Originates in the nerve sheath schwann cells. Often found in carotid artery gap, very prone to cystic or fatty necrosis. CT shows low to intermediate attenuation with intense contrast enhancement. Adjacent bone remodeling may occur.
- H. Neuroblastoma:** It is a highly malignant tumor originating from the spinal cord cell source. Often occurs in infants and young children with 95% of cases being diagnosed before 10 years of age. More than half occurs in adrenal medulla. Calcification is seen in 80-90% of cases. It tends to encase vessels and may lead to compression. Adjacent organs are displaced, may invade muscles and lymph node enlargement are often present.

Abbreviations:

CT: Computed Tomography. ENT: Ear nose and throat.

CONCLUSION

We are hereby reporting a case of a ganglioneuroma at an unusual site and with an unusual clinical presentation. The presence of neural elements and ganglion cells at this site makes it important for an inexperienced histopathologist to differentiate it from other neural lesions of the tongue, in order to avoid a misdiagnosis. An early and an appropriate recognition of such lesions can prevent the clinicians from administering inappropriate treatments. A thorough awareness on the uncommon neural lesions of the tongue can prevent any misdiagnoses and it can easily solve such diagnostic challenges.

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