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CASE REPORT

GANGLIONEUROMA PRESENTING AS A NECKMASS: A CASE REPORT

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ABSTRACT

Ganglioneuroma of the neck is a rarely reported lesion presenting as slow growing painless neck mass. We report a case of ganglioneuroma in a 7 years old female child with left-lateral neck mass slow growing over a 4-years of duration. After preliminary investigations, the patient was operated and a complete excision of the mass was performed. Post-operation biopsy confirmed a diagnosis of Ganglioneuroma that suggests that ganglioneuroma should be considered as differential diagnosis in patients with neck mass lesion.

***Key words**: Neck mass, ganglioneuroma (GN)*

INTRODUCTION

Ganglioneuromais a benign tumor that arises from the peripheral derivatives of sympathoblasts constituting cells of neural crest origin (1). Ganglioneuromas (GNs) occurring in the head and neck region are rare. As reported in literatures, only 1-5% of the affected patients present with mass lesions over the cervical area (2). The clinical picture is usually related to a mass effect, neurological dysfunctions or hypersympathetic activity due to secretary cells in the tumor (1). Imaging techniques and fine needle aspiration are supportive for the diagnosis GN, but definitive diagnosis pf GN is made after excision biopsy examination (3). Here, we present a case of GN in the cervical region, which presented as slow growing mass with symptoms of compression.

CASE REPORT

A 7 years old female child was admitted to Tikur Anbessa Specialized Teaching Hospital after she presented with a complaint of progressive left lateral painless neck swelling of 4 years duration. She had no associated symptoms except for snoring while asleep. She had no history of difficulty of breathing, dysphagia, change in voice, fever, cough, night sweating, contact with a person with chronic cough or other chronic medical illnesses.

On physical examination, the patient was healthy looking and was comfortable. Her vital signs were stable at admission. On neck examination, there was a left lateral neck mass which was 8 by 6 cm in size, hard, non-tender and mobile sideways, but not vertically. The mass was not attached to the overlying structures but was adherent to underlying structures. She did not have cervical lymphadenopathy.

Routine laboratory tests did not reveal any abnormality. Doppler Ultrasound suggested a deep cervical soft tissue mass with no vascular attachment. Computerized tomographic (CT) scan showed a well-defined, poorly enhanced. hypodense mass at the angle of the left jaw (Figure 1A &B). On fine needle aspiration cytology (FNAC), the smear showed scattered lymphoid cells as well as layers of wavy spindle cells and ganglionic cells suggestive of neural tissue origin.

Treatment: Surgery was done through left longitudinal incision over the Sternocleidomastoid Muscle. Dissection was done layer by layer till the carotid sheet was visible (Figure 2). The carotid sheet was pushed anteriorly and medially by a capsulated mass located over cervical vertebral bone. Further dissection was made below the carotid vessels and the mass was seen arising from nerve fibers. It was circumferentially separated using blunt dissection and its pedicles were ligated and transected. It was 5x7x3cm, firm, well encapsulated, white to gray in color and solid and without any infiltration (Figure 3). Histopathology showed a capsulated tissue with proliferation

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of ganglion cells admixed with tipper end spindle cells in afibrillary background with no mitosis, conclusive diagnosis being ganglioneuroma (Figure 4A & B). On the subsequent postoperative period: on 4th post operative day the patient developed fever secondary to surgical site infection with sero-sanguineous discharge. She had intermittent dry cough, dysphagia, and occasional aspiration of liquid meals. In addition, she had ptosis and myosis on left side (Horner's syndrome). There was deviation of the tongue and uvula to the contralateral side. The fever subsided following antibiotic treatment and daily wound care. But the other symptoms persisted up to a month after the surgery.

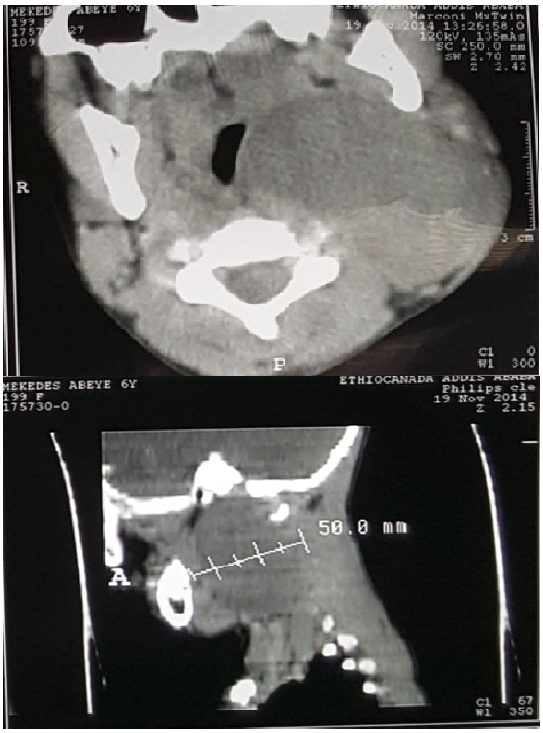

Figure 1A& B sagittal and cross sectional view of the neck mass

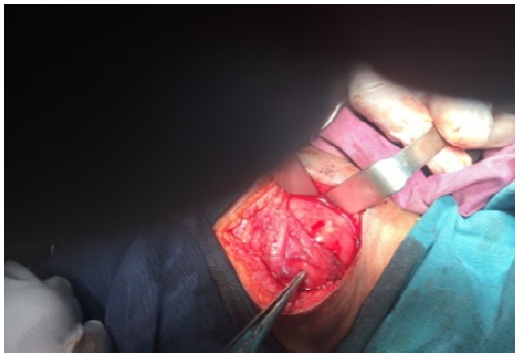

Figure 2: left neck mass posterior to carotid vessels

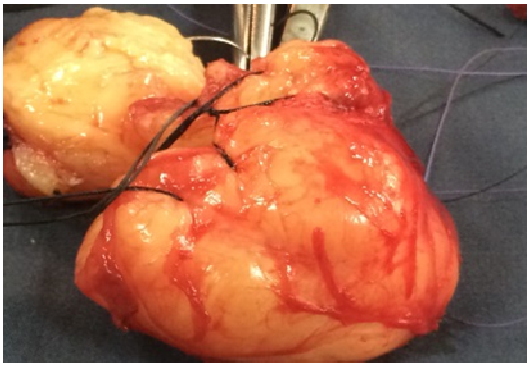

Figure 3 Macroscopicappearances of the neck mas

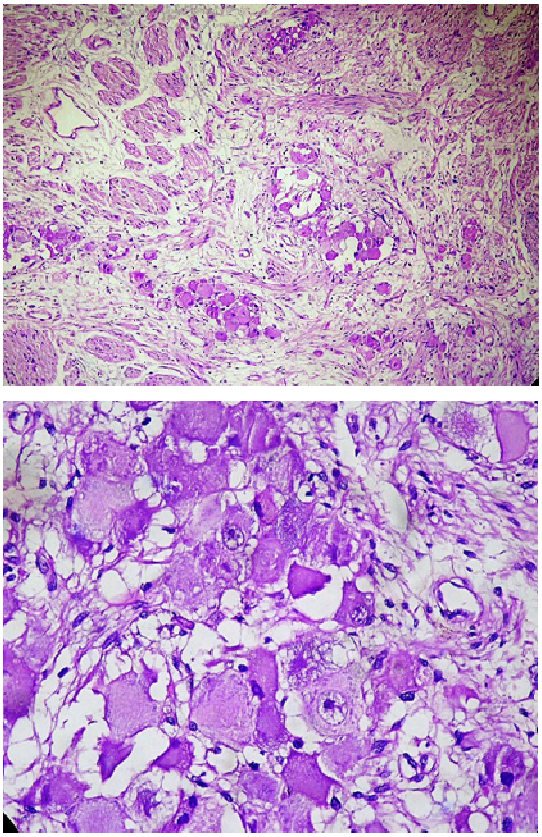

Figure 4A &B:Histology appearance of the mass (ganglion cells admixed with tipper end spindle cells in a fibrillary background with no mitosis)

DISCUSSION

GN is a benign tumor arising from sympathetic ganglion, which commonly occurs at posterior mediastinum, retroperitoneum and the adrenal gland. Rarely, it may also present in the cervical region as a slow growing painless neck mass. Neuroblastic Tumors

arise from neural crest derived cells in the sympathetic ganglia and adrenal medulla. Based on maturation and differentiation of these neural crest cells, three histologic patterns of the tumors are noted: Neuroblastoma, Ganglioneuroblastoma and Ganglioneuroma. Neuroblastomas are undifferentiated neoplasms, where as Ganglioneuroblastomas and Ganglioneuromas demonstrate evidence of differentiation (Schwannianstromal and ganglion cells) (4).

Ganglioneuromas are benign tumors that arise spontaneously or following treatment of neuroblastomas with chemotherapy or radiotherapy (5). These tumors tend to occur in older children five to seven years of age. The more aggressive forms occur below five years of age. The most frequently affected anatomical sites are the posterior mediastinum, retroperitoneum, adrenal glands. They rarely appear on head and neck soft tissues. Ganglioneuromas, unlike their malignant counterparts, tend to produce either asymptomatic mass lesions or symptoms related to compression. There are also functional ganglioneuromas that release peptides such as Vasoactive Intestinal Peptides (VIP), Somatostatins and Neuropeptide Y (NPY). These may cause some symptoms like diarrhea, sweating and hypertension. In our patient, the mass was slow growing, painless and had compressive symptoms at night but had no functional symptoms.

For perioperative diagnosis, no investigation is accurately diagnostic. Imaging modalities like ultrasound may show homogeneous, hypoechoic, well circumscribed mass. On CT: tumor size, organ of origin, tissue invasion, lymphadenopathies and presence of calcification on a well localized mass can be seen. Magnetic Resonance Imaging (MRI) may be superior to CT, yet can't discriminate benign lesions from their malignant counterparts. Currently, there are reports of ganglioneuromas diagnosed by fine-needle aspiration but definitive diagnosis was only made after surgical resection. The FNAC of our patient was suggestive of mass arising from neuronal tissue but it was not adequate enough to make the definitive diagnosis.

The treatment for this benign tumor is complete surgical resection (6). No additional adjuvant therapy is warranted. The recurrence rate after surgery is almost nil. The complication anticipated following cervical GN resection is ipsilateral Horner's syndrome due to injury to cervical sympathetic ganglion (6,7).

Conclusion: Ganglioneuroma of the neck is a rare tumor that most commonly presents as a slow growing neck mass. This tumor may be suspected in children who are otherwise asymptomatic and present with long history of enlarging neck masses. Its diagnosis can only be ascertained with postoperative pathologic examination; thus, it should be considered in all patients with neck mass. Complete surgical excision is the definitive treatment, which may lead to Horner's syndrome at times.

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