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CervicalGanglioneuroma in a Six Year Old Boy

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Abstract

Ganglioneuromas are benign neoplasm of neuroblastic origin which arises from central or peripheral parts of the autonomic nervous system. They are normally found at posterior mediastinum, retroperitoneum, and the adrenal gland but ganglioneuromas are rarely found in the cervical region. Here, we present a case of cervical ganglioneuroma in a six year old male child who presented with slow growing painless lateral neck swelling since birth.

Keywords: Children; Neck mass; Neurogenic tumor

Introduction

GanglioNeuromas (GN) are benign, neoplasm of neuroblastic origin which arises from central or peripheral parts of the autonomic nervous system. Besides, common sites such as posterior mediastinum, retroperitoneum, and the adrenal gland, GN are rarely demonstrated in the cervical region. As reported in literatures [1,2] only 1% to 5% of the patients present with neck masses.They are usually seen in children and young adults of age less than 20 years with a slight female predominance.

Case Report

A 6 year old boy presented with a slow growing mass in the left lateral side of the neck since birth slowly increasing in size especially in last two years.Mass was 7×5×3 cm,firm in consistency and was non tender.It was not associated with any systemic or compression related symptoms. There was no relevant medical,family and personal history.With provisional diagnosis of tubercular lymphadenitis/ lymphoma,two cervical lymph nodes were excised and sent for excisional biopsy to render the diagnosis,largest measuring 3×2×2 cm and smallest being 3×1.5×1 cm.On cut surface,they were pale white and firm in consistency.Onmicroscopy,it showed well encapsulated tissue bit containing few scattered mature ganglion cells surrounded by schwanian rich stroma suggesting Ganglioneuroma-Schwanian dominant neuroblastictumor. Further on IHC(ImmunoHisto Chemistry),the section was positive for various IHC markers like S-100,Calretinin,Synaptophysin,CGA,Ki-67 suggesting mature ganglioneuroma.Radiologically,a contrast enhanced MRI was done which showed a large 6.5×4.5 cm T1W hypo and T2W hyperintense mass lesion seen at left carotid space region and extending superiorly in to parapharyngeal space and inferiorly upto cricoid cartilage displacing the neck vessels(carotid and jugular vein) antero laterally.Lesion showed intense enhancement after contrast administration.In addition multiple variable sized cervical lymph nodes were seen on left side.Features were suggestive of Ganglioneuroma with cervical lymphadenopathy.Then surgical excision was planned.

Under general anaesthesia,a transverse incision was given over the swelling for the removal of tumor. The internal jugular vein and the common carotid artery which were displaced by the tumor were retracted and preserved. The excised lesion was well circumscribed, making an intact removal possible. The patient was extubated immediately after the surgery with no clinical signs of respiratory distress.The patient was well and without any complaints in the post-operative period.

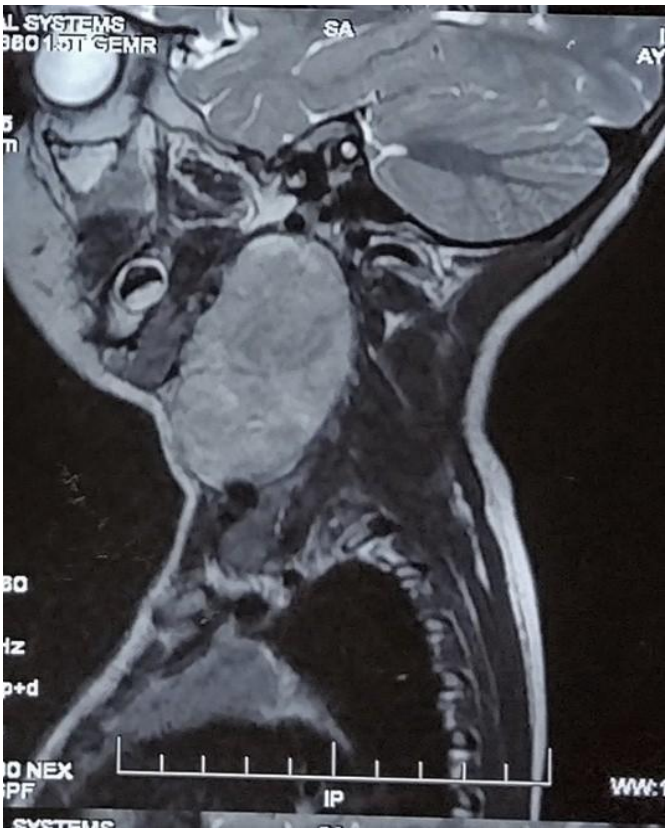

Figure 1:MRI (T2) saggital section shows a mass of 65×45 mm centered in the left carotid space, extending superiorly to the parapharyngeal space and inferiorly uptocricoids cartilage.

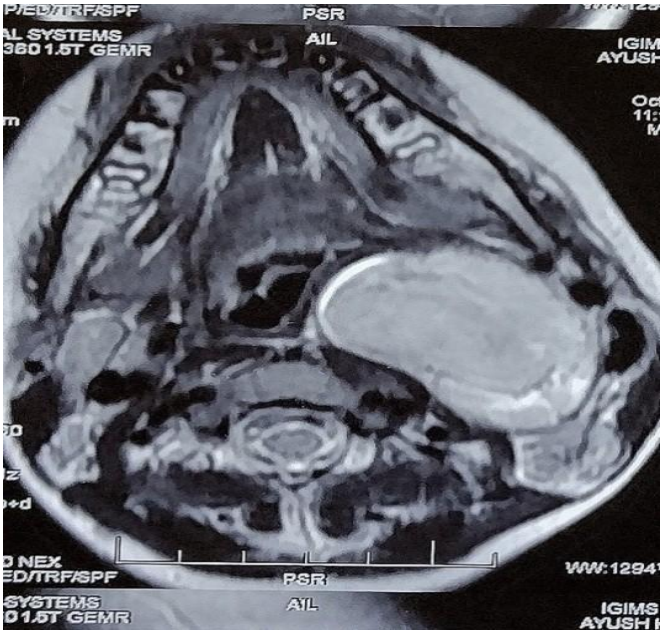

Figure 2:MRI (T2) axial section shows a mass of 65×45 mm centered in the left carotid space, extending superiorly to the parapharyngeal space and inferiorly uptocricoids cartilage.

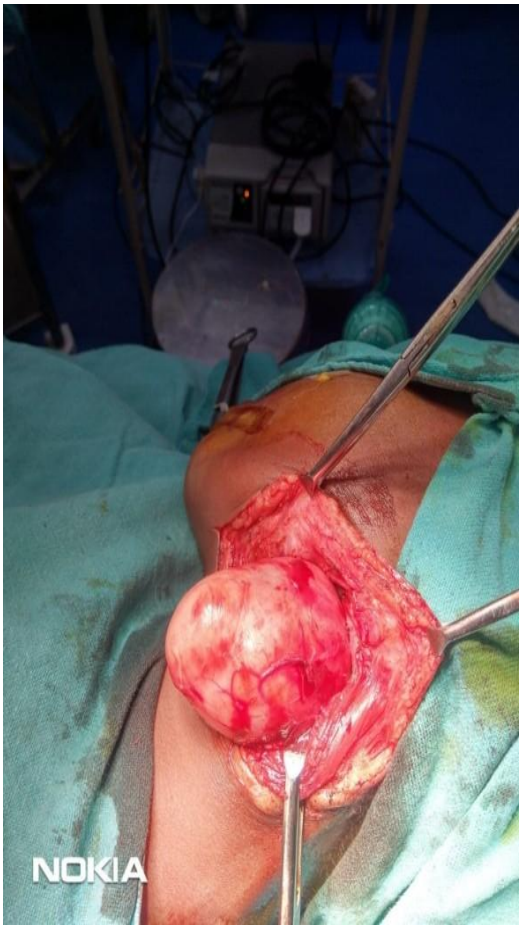

Figure 3:A well circumscribed mass before the excision.

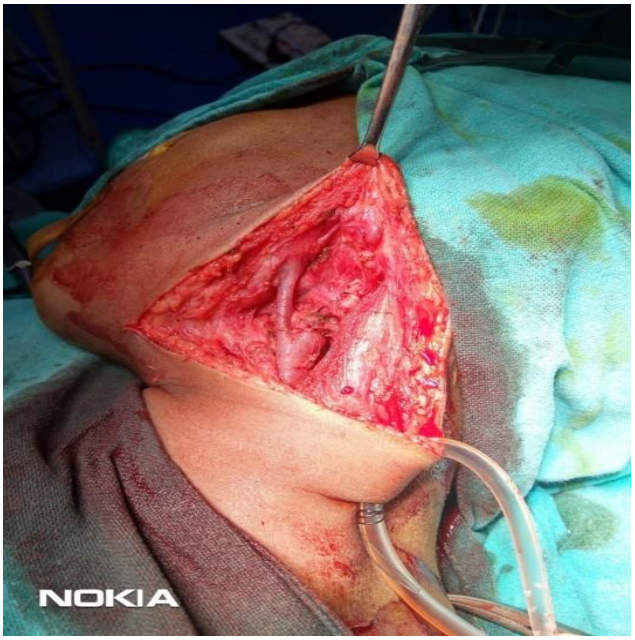

Figure 4: After excision of mass-showing internal jugular vein.

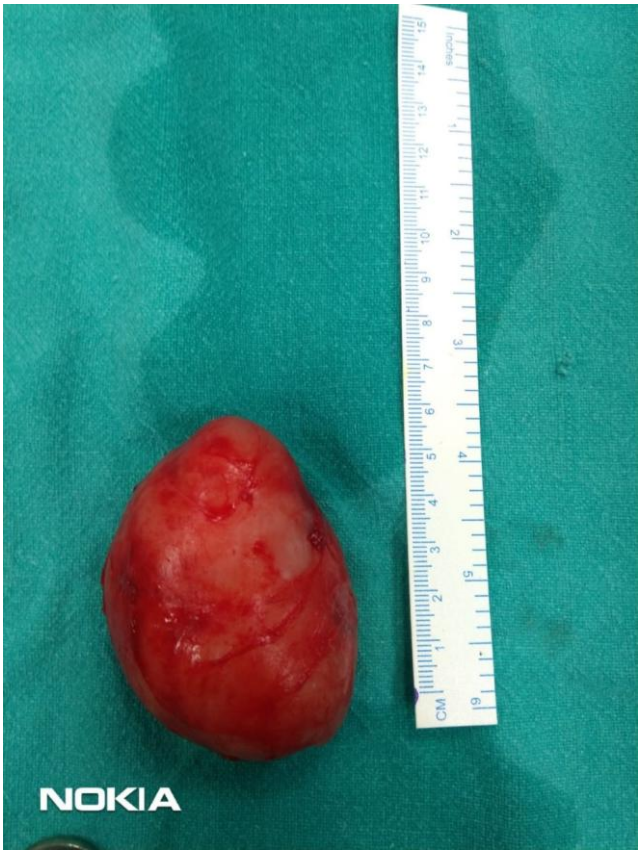

Figure 5: Surgical Specimen

Discussion

Ganglioneuroma was first described by Loretz in 1870 [3]. It originates from the primordial neural crest cells which are undifferentiated cells of the central nervous system. These cells are

present in the sympathetic ganglia and adrenal medulla, indicating that these tumors may grow nearly anywhere along their distribution. Ganglioneuroma usually occurs in the abdomen or thorax, but it can also occur in the head and neck region, with the latter being it's least common location. Actually, this is one of the rarest neck tumors. The first report of a ganglioneuroma located in the neck was made in 1899 by de Quervain [4] .The most frequent origin in the neck region is the cervical sympathetic chain, but it can also be located in the larynx, pharynx, and ganglion nodosum of the vagus nerve [5].

These types of tumors are more common at a young age, as 60% of patients with ganglioneuroma are <20 yrs old 10. The average age of presentation is 11 years old, with a slight preponderance in the female gender [6]. They are usually slow-growing masses, with most of them being silent in the early stages of development as in our case [5].

The reviewed literature suggests the possibility of these tumors being secretory [3, 5-7]. However, our case and the ones reviewed were not metabolically active. Nevertheless, the differential diagnosis between ganglioneuroma and neuroendocrine tumors can be quite difficult. According to the North American Neuroendocrine Tumor Society guidelines(NANETS), the initial testing for pheochromocytoma or paraganglioma must include measurements of fractionated metanephrines in plasma, urine, or both, as available. Given their high sensitivity for neuroendocrine tumors (reaching 100% in children and 99% in adults), they are part of the initial routine evaluation of this kind of masses. In other case series, fine-needle aspiration failed to identify this kind of tumor in 60% of cases. Nevertheless, it can be of tremendous value in the differential diagnosis in pediatric patients, as it is a rapid, safe and cost-effective method, although sedation is often required.

In addition, imaging is crucial not only on the differential diagnosis of the neck mass but also to plan surgery. Generally, both Computed Tomography (CT) and MRI are mandatory. CT frequently reveals a well-defined mass, which displaces the surrounding structures. MRI shows intermediate signal intensity and cystic or non-homogeneous contrast enhancement. On the other hand, functional imaging with Meta-iodobenzylguanidinescintigraphy has a high specificity for neuroendocrine tumors and can aid in the precise localization of that kind of disease. Several authors report having used it preoperatively for differential diagnostic purposes. This examination was not used in our patient as the by-products of sympathetic response were negative. As in our case, preoperative diagnosis is not possible most of the times.

Surgical resection of these tumors is the gold standard treatment[8-10]. The approach may vary. As in our case, a transcervical approach can be used; however, transoral, transparotid, transcervical–transpharyngeal, and infratemporal fossa approach can also be used depending on the location, size, and pathological type. Mandibulotomy has also been performed in a few cases for better exposure of the skull base. Damage to the nearby neural and vascular structures during surgery may result in significant morbidity.However; these symptoms tend to resolve rapidly.

Definitive diagnosis is based on histopathological analysis [9]. The International Neuroblastoma Pathology Classification defines four categories of peripheral neuroblastic tumors characterized by the grade of neuroblastic differentiation and the degree of Schwannian stromal development. Neuroblastomas are cellular neuroblastic tumors without prominent Schwannianstroma. Ganglioneuroblastomas are subdivided into intermixed and nodular categories. The former comprises neuroblastic elements in an abundant Schwannianstroma, and the latter is a composite tumor with a neuroblastic nodular component and either an intermixed ganglioneuroblastoma or a ganglioneuroma component. As in our case, ganglioneuromas represent the most differentiated form, consisting of mature ganglion cells distributed in a predominant Schwannianstroma. Ganglion cells vary in distribution and number and can be either localized or widely scattered. Schwann cells may ensheathneuritic processes or may be arranged in small intersecting fascicles, which are separated by loose myxoidstroma. Neuroblasts should be negative in order to make this diagnosis. Furthermore, significant atypia, mitoses, or necrosis must be absent [10].

If the diagnosis is confirmed after surgical excision, it can be curative. The prognosis is favorable, as ganglioneuromas do not have metastatic potential [11]. The patient does not require further treatment. Even in the case of incomplete resection, it does not have the potential to recur. Radiation should be avoided in these cases, especially in young ages, as it can cause growth retardation and later problems. On the other hand, other categories of neurogenic tumor have different behaviors. Neuroblastoma is a malignant solid tumor, and ganglioneuroblastoma has moderate malignant potential, which may require the need for close follow-up and for complimentary treatment, such as chemo- or radiotherapy.

Conclusion

Ganglioneuroma of the cervical region is a rare benign soft tissue neoplasm of neuroblastic origin of neural crest cells. Surgical excision is the only definite treatment of cervical ganglioneuroma to prevent further growth and compression of the neighboring structures. It is also the only way to confirm the diagnosis. These tumors are usually not aggressive. Injury during surgery may result in significant morbidity.

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