

Cervical Sympathetic chain ganglioneuroma : case report and review of literature

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ABSTRACT: Ganglioneuroms are benign tumor of the autonomic nerve fibers, arising from neural crest and the reported incidence of ganglioneuroma is one per million population. They usually present in patients under 20 years of age with a slight female predominance .The most common localization is the posterior medistinum followed by the adrenal gland ,retroperitoneum (sympathetic ganglia) ,and head& neck.In the neck cervical sympathetic chain is the most frequent site of origin . Cervical Sympathetic chain ganglioneuromas are usually asymptomatic neck masses and complete surgical resection is the treatment of choice. Surgical excision via a cervical approach offers definitive therapy but may be associated with an iatrogenic Horner's syndrome for which the patients should be counseled prior to operative procedure . Being quite rare among other neurogenic tumors, A case of cervical Sympathetic chain ganglioneuroma is presented and the literature is reviewed.

KEY WORDS- Cervical ganglioneuroma, Head and neck tumors, neurogenic neck tumours , sympathetic chain.

I. INTRODUCTION:

Ganglionuroma occurring in the neck are uncommon. Shumacker and Lawrence (1939) state that Cervical Sympathetic chain ganglioneuromas is one of the rarest of neck tumors. Cervical sympathetic chain is the most frequent structure of origin in the neck. Other sites of origin include the larynx, pharynx and ganglion nodosum of the vagus nerve. Ganglioneuromas can be found in the central nervous system or peripherally in the sympathetic system Unusual sites include the spermatic cord, heart, bone, and intestine [2]It is a slow growing tumour with so far no recorded metastatic potential. Surgical excision is the treatment of choice. We present a case of cervical sympathetic chain ganglioneuroma discussing the diagnosis, treatment protocols and postoperative complications of this benign tumor.

CASE REPORT : A 38 years old female presentd with diffuse fullness on the right side neck for last 5 years. (Fig 1). Examination revealed a lobulated, painless, immobile mass of 10cm x 5 cm size with normal overlying skin . The displacement of the carotids was so anterior that it can be seen pulsating and appeared quite sinister. Fine needle aspiration cytology (FNAC) showed a preponderance cluster of mature ganglion cells and mixture of spindle Schwann cells. A diagnosis of ganglioneuroma was suggested Ultrasonography was done to note a round, homogeneous, mixed lesion . Magnetic resonance imaging (MRI) depicted a well defined spindle mass lesion of 9 cm x 4 cm originating from the carotid sheath, encroaching upon the right parapharyngeal spaces, displacing the common carotid and the external carotid anteriorly and causing compression of adjacent soft tissue and vascular structure suggested a neurogenic tumor. Abdominal ultrasonography (USG) showed no abnormality and urinary vanillylmandelic acid (VMA) levels were normal Patient was planned for surgical excision. Neck exploration was carried out, The dissection was rendered tedious due to the superficial lying great vessels and a compressed internal jugular veins. The vagus nerve identified and preserved along with the vessels and it was retracted anterior and medially to define a fusiform lesion arising from the sympathetic chain , when traced upward towards the neck a stump of 3 cm of the nerve was identified (Fig 2). The surgical removal of the lesion was however done with ease, owing to the well encapsulated nature of the lesions, an intracapsular dissection could not be feasible and wound was closed after provision of surgical drain . On macroscopy, it was a thick-walled, tangled, pedicled mass 12 cm × 15 cm in sizeSubmitted histopathology, showed numerous mature ganglion cells with eccentrically placed nucleus in background of Schwann cell proliferation .Histological appearance of more than 50% presence of Schwann cell population along with

neuroblastic cells confirms the diagnosis (Fig3) . Within the first day of the operation, the patient developed Horner syndrome, characterized by myosis and ptosis. She had also difficulty in swallowing her saliva. One month after the operation, ptosis and difficulty in swallowing recovered, but myosis persisted. The tumor did not show any signs of regrowth or recurrence within 30 months after the operation.

II. DISCUSSION AND REVIEW OF LITERATURE

Medical literature is replete with noting on rarity of ganglioneuroma. In 1939, Schumacker and Lawrence¹ commented that cervical sympathetic chain ganglioneuroma was indeed the rarest neck tumours. Elsewhere in the neck region they arise from the larynx, pharynx, and the hypoglossal nerve, along the length of the vagus and the intervertebral foramina and thereby extending to even the spinal cord. It can also manifest in the thoracic cavity (posterior mediastinum), abdominal cavity (retro-peritoneum, adrenals and pelvic sympathetic ganglia) as well as in the orbit. The primordial tissue of sympathetic nervous system, is the seat of origin of neuroblastic neoplasm which is neural crest and neural tube derived. as per the Shimada classification, The family of neuroblastic neoplasm includes three histological variants; neuroblastoma, ganglioneuroblastoma and ganglioneuroma, During the pathological progression, this neoplasm shows different degree of maturation with complete immature cells in neuroblastoma, the ganglioneuroblastoma and ganglioneuroma follows next in line, depending on degree of maturation². The first two are the malignant spectrum of this neoplasm. However the recent International Neuroblastoma Pathology Committee³ has in histological detail to classify them in four categories viz the

- i. neuroblastoma,
- ii. neuroblastoma intermixed with ganglioneuroblastoma,
- iii. nodular ganglioneuroblastoma and
- iv. ganglioneuroma.

This is the modification of Shimada's classification. The same classification further sub-divides ganglioneuroma into matured and immature types depending upon the percentage of presence of Schwann cells and neuroblastic cells. Thus the ganglioneuroma is a well differentiated neoplasm of the younger aged population between 2-15 years, with more neural tissue and scanty Schwann element and no chromaffin element. It is confusing to know the differences in neurogenic tumours till we understand them according to their origin. Nerves consist of neural tissue with supporting tissue like schwann and closely connected paraganglia. Neurogenic tumours may arise from neural cells itself as in the case of ganglioneuroma and its subtypes, or from paraganglion cells, like carotid body tumours and paragangliomas, and lastly from the supporting tissue like Schwann cells which comprises of schwannoma.. The pathology may not ring a bell in the doctor's mind due to its rarity in presentation. The gross suspicion of this neoplasm is made when it presents with either compression symptoms or a large mass, which are generally late. Autonomic dysfunction like diarrhoea, profuse sweating, virilisation, hypertension and alopecia are attributed to immature neuroblastic tumours. These autonomic dysfunctions as a result of catecholamine secretion occur in 37% of these cases. They are slow growing tumour with so far no recorded metastatic potential and complete excision being suffice to bring about a cure. Ganglioneuroma should be distinguished from the immature forms like neuroblastomas and neurofibroma. Histological appearance of more than 50% presence of Schwann cell population along with neuroblastic cells confirms the diagnosis. The metaiodobenzylguanidine scan (MIBG) is trusted to show 88% sensitivity and 99% specificity for these tumours along with carcinoid and pheochromocytoma. Immunohistochemistry, in which ganglion cells stain for neuron specific enolase (NSE) and Schwann cells stain for S-100 protein⁵, Useful to identify the biochemical and immunology of cell by using antigen and visible labelled tagged antibodies. Computed tomography (CT) scanning is a preferred methods for imaging ganglioneuromas and ganglioneuroblastoma^{6,7,8}. MRI is the modality of choice for evaluating the extension of tumors.⁹. and provides important pre-operative information for planning optimal surgical treatment. Immunohistochemistry is important to identify the biochemical and immunology of cell by using antigen and visible labelled tagged antibodies, to differentiate specific cellular components occur in 37% of these cases which secrete vanillylmandelic acid and homovanillic acid and the presence of tumour on sites as ganglia and retro-peritoneal adrenals⁴



Fig 1:

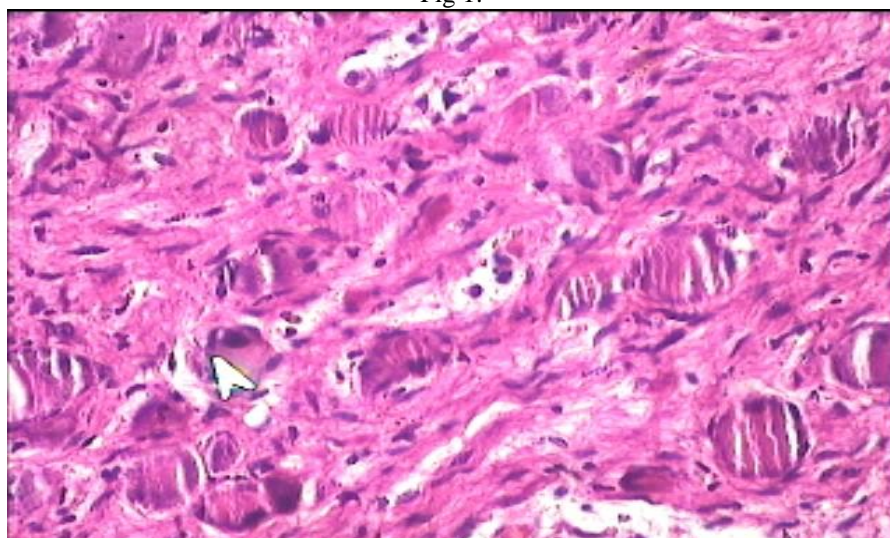


Fig 2- Histopathology of ganglioneuroma composed of mature ganglion cells in Schwannian stroma (H.E. 440×).

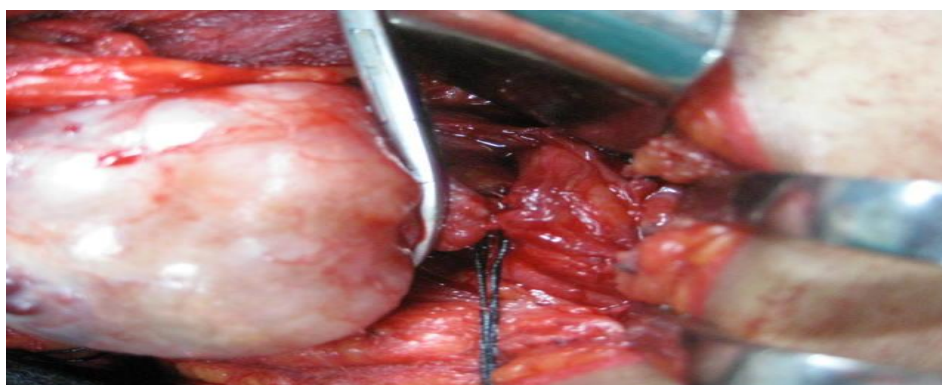


Fig 3 - Intraoperative view of the mass with its distal edematous end continuous with neural components of the neck.

III. CONCLUSION

Neuroblastic tumors of the head and neck, namely, neuroblastoma, ganglioneuroblastoma and ganglioneuroma are rare entities, accounting for 6% of the tumors of childhood. Ganglioneuromas are most frequently diagnosed in patients between the ages of 10 and 29 years. and are most commonly located in the posterior mediastinum followed by the retroperitoneum. They are usually located in the abdomen (65–80%), or thorax (10–15%), and rarely in the neck (5%) [3]. These tumors define a spectrum of sympathetic neuroectodermal tumors, ranging from the undifferentiated neuroblastoma to the mature ganglioneuroma. The presence of immature tissue in neuroblastoma and ganglioneuroblastoma indicates malignant or potentially malignant behavior; ganglioneuroma is composed entirely of ganglion cells and Schwannian stroma, which is considered benign [4]. A case of ganglioneuroma was first reported by Loretz in 1870 and ganglioneuroma of the neck was first reported by De Quervain in 1899 [1]. Ganglioneuroma arises under the age of 20 years in 60% of the cases, the median age of presentation is approximately 7 years with a slight preponderance of cases in

females [1], [4]. Compared to other neuroblastic tumors, ganglioneuroma occurs in older patients. The ratio of neuroblastoma to ganglioneuroma is approximately 6:1 to 10:1 [2]. Ganglioneuroma most often manifests as an asymptomatic mass. Clinically, signs and symptoms of cervical ganglioneuromas are usually related to compression of vital structures such as nerves or vessels of the neck. The tumors involving the cervical sympathetic chain causes Horner syndrome characterized by ptosis, myosis, ipsilateral facial anhidrosis and flushing. In 37% of cases, catecholamines secreted by the tumor may increase levels of VMA or homovanillic acid (HMA) in the plasma and urine, which may lead to hypertension, diarrhea, sweating, flushing and renal acidosis [1], [5]. We did not detect an elevation of hormonal levels in our patient. In I131-MIBG scintigraphy an increased uptake in the tumor tissue can be detected [4]. Most characteristic histologic feature of ganglioneuromas is the presence of mature ganglion cells. However in 25% of ganglioneuromas elements of immature neurogenic tumors can be seen. It is hypothesized that neuroblastic tumors undergo a maturational process or spontaneous regression of neuroblastoma to ganglioneuroma [1]. Macroscopically, ganglioneuroma may appear encapsulated, although a true capsule is infrequent [2]. The cervical sympathetic chain is the most frequent structure of origin in the neck. Other sites of origin are larynx, pharynx and ganglion nodosum of the vagus nerve [2]. Apart from neuroblastic tumors, there are other rare tumors of the neck. Ganglioneuroma may be mistaken for such tumors as paraganglioma, oncocytoma, lipoma, plasmocytoma, embryonic rhabdomyosarcoma, Ewing's sarcoma and non-Hodgkin lymphoma and such infections as actinomycosis, toxoplasmosis, tuberculosis, infectious cervical lymphadenitis and branchial cleft cyst [3], [6]. USG reveals a homogeneous, hypoechoic mass with well-defined borders however MRI and computerized tomography (CT) are more valuable imaging techniques in the diagnosis of ganglioneuroma. MRI is superior to CT in the diagnosis of intraspinal tumors [7]. The tumor has been shown to have a capsule and calcifications both radiologically and histologically [5], [8]. Although ganglioneuroma tends to be a more homogeneous tumor than neuroblastoma or ganglioneuroblastoma, it is not possible at imaging evaluation to discriminate among these three tumors [2]. There are rare reports of metastatic ganglioneuroma. It is believed that these tumors represent metastases of neuroblastoma or ganglioneuroblastoma that have subsequently matured to ganglioneuroma; these patients have an excellent prognosis [4].

Surgical excision is the treatment of choice to confirm the diagnosis and prevent further growth and compression of neighboring structures. These tumors are not aggressive, and even in the case of incomplete resection, residual ganglioneuroma will not regrow or produce symptoms [3]. Significant morbidity may occur as a result of intraoperative sacrifice of neural structures and vasculature associated with the tumor. Horner Syndrome and significant aspiration of thin liquids are frequent complications. However these symptoms usually resolve in the postoperative follow-up [1]. Chemotherapy and bone marrow transplantation are also used in the treatment [2]. Radiotherapy should be avoided in children as it increases the risk of growth retardation and tumor development secondary to radiation [3]. Atives in this above mentioned case report. 2They are usually located in the abdomen (65–80%), or thorax (10–15%),.

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