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RESEARCH ARTICLE

AN UNUSUAL NECK SWELLING: MALIGNANT GANGLIONEUROMA, A RARE ENTITY.

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Abstract

Introduction: Ganglioneuroma (GN) is a rare, benign, non-invasive and neurogenic tumour. It has a neuroectodermic origin and is localized along the sympathetic trunk. Due to its rarity and the lack of specific signs and symptoms it is often difficult to reach definite diagnosis prior to surgical exploration and complete surgical excision and pathological examination. **Material and Method:** A 6 year old female presenting with swelling in the right side of neck associated with difficulty in breathing to the out-patient department of Cardiothoracic Surgery of Dhiraj Hospital was selected. **Result and Analysis:** The patient was discharged on post-operative day 9 with stable vitals and no post-operative complications. **Discussion:** It has a higher female predominance and commonly presents as an enlarging neck mass. In the neck due to proximity to the thyroid gland it occasionally presents with signs of Horner's syndrome. Though usually asymptomatic, they may sometimes cause symptoms when vital structures are compressed or when there are high levels of catecholamine. MRI and CT are the mainstay of diagnosis for such tumors. Malignant ganglioneuromas are rare. Surgical excision of ganglioneuromas is the mainstay of the treatment.

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Introduction:-

Ganglioneuroma of the neck is a rare tumor usually seen in children and young adults less than 20 years of age. They arise from sympathetic system and it's benign in nature. It has a higher female predominance and commonly presents as an enlarging neck mass. In the neck due to proximity to the thyroid gland it occasionally presents with signs of Horner's syndrome. It also causes compression of vital structures in the neck. Clinically and radiologically these lesions can be mistaken as something else. Common sites are adrenal gland, retroperitoneum, mediastinum and rarely neck (8%). We report a case of mediastinal ganglioneuroma extending into the neck in a 6 years old female child.

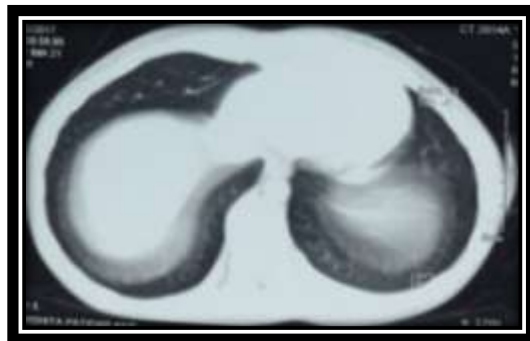
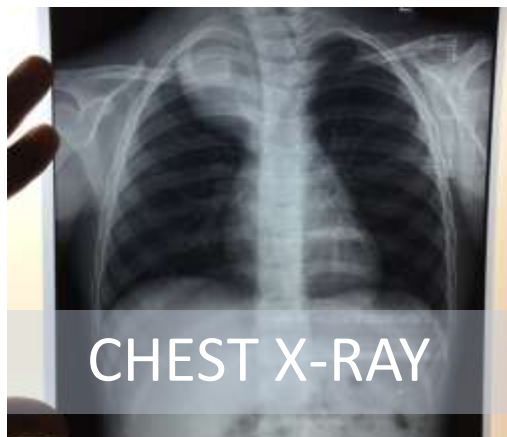
Case Report:-

A 6 years old female child presented with a right side neck swelling since 1 year. The swelling was gradually increasing in size, painless and associated with difficulty in breathing. There was no associated dysphagia or fever. On local examination it was approximately about 6cm x 4cm swelling in right supraclavicular region and was non-

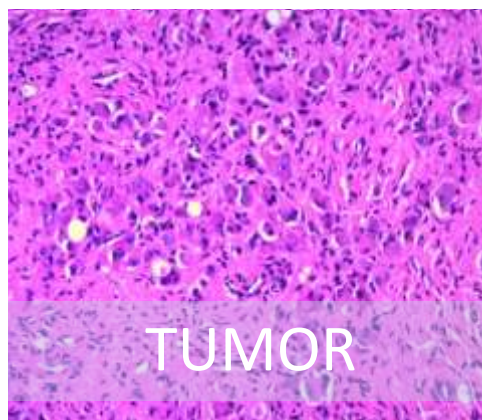
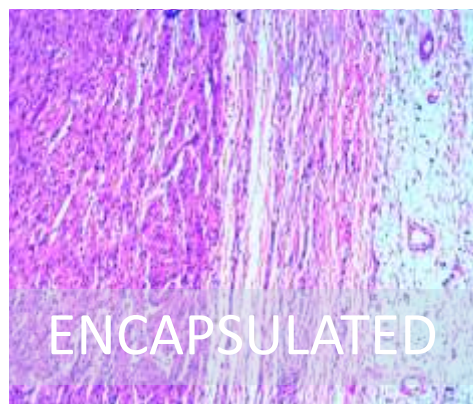
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tender, mobile but not moving with protrusion of tongue. FNAC result showed hypocellularity with occasional pleomorphic cells with prominent nucleoli with enlarged nucleus and abundant cytoplasm suggestive of malignant tumor. USG suggested ill-defined, heterogenous, predominantly hyperechoic solid mass lesion with internal macro-calcification and subtle areas of necrosis and minimal internal vascularity of size 6.1 x 4.1 x 2.8 cm in right supraclavicular region and extending in right infra-clavicular region displacing trachea to left side. The lesion also displaced and compressed the adjacent carotid vessels. CT scan of Neck and Thorax suggests a large mild enhancing soft tissue mass lesion measuring 7.5 x 7 x 7 cm in right side superior mediastinum with supra clavicular extension abutting and displacing trachea to left side. It also abuts right common carotid artery, subclavian artery and vertebral artery without significant luminal narrowing. It compresses internal jugular vein with complete luminal occlusion. There is no evidence of necrosis, calcification or cystic area. During surgery (In Total Excision) there was a large 20 x 10 cm dumbbell shaped mass in right supraclavicular region extending in the neck, behind clavicle in right thoracic region abutting the right lung, right CCA, right IJV, right subclavian artery and vertebral artery completely engulfed by tumor. Posterior portion of tumor engulfed multiple nerve roots with multiple scalene group of lymph nodes and large retro aortic lymph node. Histology was suggestive of Ganglioneuroma (Schwannian Stroma-Dominant) category, Maturing subtype, Stage IIB. Ipsilateral regional lymph nodes: single scalene lymph node shows presence of tumor. Other lymph nodes and thymus shows no evidence of tumor. Procedure was un-eventful with an uneventful post-operative course. Patient was discharged on post-operative day 9 and is doing well on follow-up.



CT SCAN OF NECK AND
THORAX SHOWING
THE MASS



Discussion:-

Loretz first reported ganglioneuroma in 1870 while De Quervain in 1899 first reported ganglioneuroma of neck [1]. Cervical ganglioneuroma, benign tumors of neurogenic origin, are rare and account for 6% of childhood tumors [2]. They arise from cervical sympathetic chain [3]. 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. Ganglion cells, Schwann and stroma are the component of such tumors [4]. Though usually asymptomatic, they may sometimes cause symptoms when vital structures are compressed or when there are high levels of catecholamines. Increase in catecholamine level lead to rise in levels of VMA and HMA in plasma or urine resulting in hypertension, diarrhoea, flushing, sweating, renal acidosis and other symptoms of catecholamine excess [5]. In a case report by Junli, there was only one case of multiple ganglioneuroma on one side of neck while 3 cases with bilateral neck masses over a period of 10 years. MRI and CT are the mainstay of diagnosis for such tumors. Such tumors have capsule and calcifications. Radiologically it is impossible to differentiate between ganglioneuroma, neuroblastoma and ganglioneuroblastoma. On FNAC the characteristic finding of ganglioneuroma

is presence of mature ganglion cells but in 25% of the cases immature neurogenic tumors can be seen. Macroscopically they appear encapsulated but true capsule is infrequent. 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 pheochromacytoma. Immunohistochemistry, in which ganglion cells stain for neuron specific enolase (NSE) and Schwann cells stain for S-100 protein [6]. Usually malignant ganglioneuromas are rare. They represent metastases of neuroblastoma or ganglioneuroblastoma which have later matured to ganglioneuroma. Such patients have excellent prognosis. Our case is that of malignant ganglioneuroma.

Peripheral neuroblastic tumors represent a spectrum of diseases from undifferentiated and malignant NB to well-differentiated and benign GN [7].

International neuroblastoma pathology classification (the Shimada System)

1. NB (schwannian stroma-poor)
2. Intermixed GNB (schwannian stroma-rich)
3. Nodular GNB (schwannian stroma-rich/stroma-dominant and stroma-poor)
4. GN (schwannian stroma-dominant),
5. Maturing
6. Mature.

Surgical excision of ganglioneuromas is the mainstay of the treatment. These tumors being non aggressive donot recur and residual compnents of the tumors also shall not cause any symptoms. Complication from surgery for cervical ganglioneuroma is the injury to compressed vascular and neural structures mostly resulting into Horner's syndrome, but its symptoms resolve soon. Chemotherapy and bone marrow transplantation are also used in the treatment [8]. Radiotherapy should be avoided in children as it increases the risk of growth retardation and tumor development secondary to radiation [9].

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