Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 7, Issue, 04, pp.15195-15197, April, 2015

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

ISSN: 0975-833X

CASE REPORT

SYMPATHETIC CHAIN GANGLIONEUROMA - A CASE REPORT

[1,*]Sushil Gaur, [2] Sunil Kumar Singh Bhadouriya, [3] Vivek Kumar Pathak and [4] Mamta Bisht

Dept. E.N.T and Head and Neck Surgery, School of Medical Science and Research, Greater Noida, India

ARTICLE INFO

Article History:
Received 20th January, 2015
Received in revised form
07th February, 2015
Accepted 23rd March, 2015
Published online 30th April, 2015

Key words:

Ganglioneuroma,
Sympathetic chain,
Neoplasm,
Cervical.

ABSTRACT

Ganglioneuromas are extremely rare. Ganglioneuroma, of the parapharyngeal space of the neck was surgically operated in a 7 year old boy, presenting with a 5 year history of diffuse neck swelling. Sympathetic chain neck ganglioneuroma presents infrequently. The FNAC finding of mature ganglion interspersed with Schwann cells is also rare. It needs to differentiate ganglioneuroma from the rest of the tumours of primordial neuroblastic tissue. Although very rare, ganglioneuromas must be considered in the differential diagnosis of neck masses because pathology may not ring a bell in the doctor's mind due to its rarity in presentation.

INTRODUCTION

Tumors of the sympathetic ganglia include ganglioneuromas, ganglioneuroblastomas, and neuroblastomas (Albonico *et al*., 2001) Ganglioneuroma can be found anywhere along the sympathetic chain, but is most commonly located in the posterior mediastinum and retro peritoneum (Enzinger *et al*., 1995; George *et al*., 2001). The neck is a rare site of presentation (Weber *et al*., 2000; Weiss *et al*., 2008). Ganglioneuroma is the most differentiated benign counterpart of neuroblastoma and originates similarly from neural crest cells (Weber *et al*., 2000; Califano *et al*., 2001). Ganglioneuromas are encapsulated tumors, composed of large ganglion cells, nerve sheath cells, and nerve fibers. These tumors occur predominantly in infants and children.The signs and symptoms of cervical ganglioneuromas are usually related to the mass effect and nerve dysfunction, but these tumours often present as swelling with no specific symptomatology, as in this present case (Califano *et al*., 2001). Here, we report a case of ganglioneuroma that was found in an unusual location the parapharyngeal space (Albonico *et al*., 2001; Enzinger *et al*., 1995). Complete excision was possible by cervical approach.

CASE REPORT

A 7 year old boy presented with fullness in the right neck region noted at 2 years of age and the pathology growing over the years.

Corresponding author: Sushil Gaur
Dept. E.N.T and Head and Neck Surgery, School of Medical Science and Research, Greater Noida, India

Preliminary investigation of CT scan depicted a mass lesion of 9cm x 4 cm encroaching upon the right parapharyngeal space displacing the common carotid and the external carotid anteriorly. Examination confirmed the displacement of the carotids so anterior so as to be felt and seen in the subcutaneous tissue, pulsating and appearing quite sinister, the sternomastoid was stretched and the anterior border could be palpated laterally with an intervening space through which the pathology of the parapharyngeal space could be felt. Fine needle cytology showed a cluster of mature ganglion cells and mixture of spindle Schwann cells.

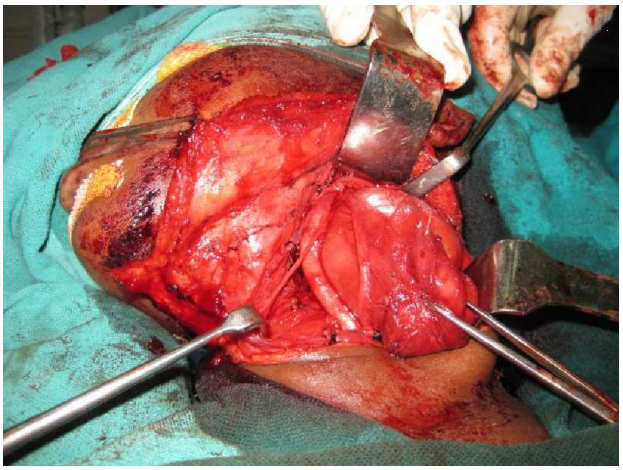

Figure 1. Intraoperative photograph of parapharyngeal space ganglioneuroma

Provisionally diagnosed as ganglioneuroma later diagnosis was confirmed with the use of immunohistochemistry. Ultrasonography was done to know the relation of the mass to major blood vessels and consistency. Neck exploration was carried out. Dissection was tedious due to the superficial lying great vessels and a compressed internal jugular vein. The vagus nerve identified and preserved. The Vessels were retracted anterior and medially to expose the mass arising from the sympathetic chain (Fig-1). The surgical removal of the mass was done and wound was closed in two layers, after fixing of surgical drain. Excised Tissue was sent for histopathological examination which showed Schwann cells, fibrous tissue and embedded within were large cells with abundant cytoplasm, large nuclei and prominent nucleoli (ganglion cells). There was no evidence of atypia / mitosis / necrosis. (Fig-2)

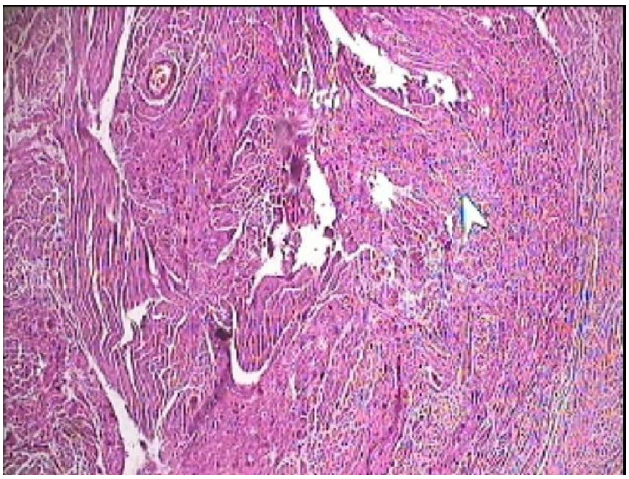

Figure 2.

Based on the histopathological features a diagnosis of ganglioneuroma was made. Post- operative period was uneventful; patient was discharged on 8th postoperative day.

DISCUSSION

Medical literature is replete with noting on rarity of ganglioneuroma. In 1939, Schumacker and Lawrence commented that cervical sympathetic chain ganglioneuroma was indeed the rarest neck tumour (Schumacker and Lawrence, 1939). They have been described in the neck region to 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 manifest in the thoracic cavity (posterior mediastinum), abdominal cavity (retro-peritoneum, adrenals and pelvic sympathetic ganglia) as well as the orbit. The primordial precursor tissue of sympathetic nervous system (neural crest and neural tube derived), is the seat of origin of neuroblastic neoplasm.

The family of neuroblastic neoplasm includes entries of three histological variants; neuroblastoma, ganglio-neuroblastoma and ganglioneuroma, as per the Shimada classification. Somewhere during the pathological progression of this neoplasm different degree of maturation may occur and may render the neuroblastic tissue with a histological appearance of complete immature cells as in neuroblastoma, with the ganglio-neuroblastoma and ganglioneuroma to follow next in line, depending on degree of maturation (Bove & Mc Adams, 1981). The first two are the malignant spectrum of this neoplasm. However the recent International Neuroblastoma Pathology Committee (Shimada *et al*., 1999) classified them in four categories viz. the 1) neuroblastoma, 2) neuroblastoma intermixed with ganglio-neuroblastoma, 3) nodular ganglio-neuroblastoma and 4) the ganglioneuroma. This is the modification of Shimada's classification (George *et al*. 2001). The same classification further sub-divides ganglioneuroma into mature 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 population aged between 2-15 years, a slight variation in age being reflected in many studies. The sex incidence is agreed to be slightly more common in girls than boys.

The gross suspicion of this neoplasm is made when it produces compression symptoms which are generally late, or presents as a mediastinal shadow or presents as a peritoneal occupying lesion. The pathology may not ring a bell in the doctor's mind due to its rarity in presentation. Another much easier presentation may follow the compressive symptom along with autonomic dysfunction like diarrhoea, profuse sweating, virilization, hypertension (George *et al*., 2001) and alopecia, but these are attributed with immature neuroblastic tumors which secrete vanillyl mandelic acid and homo vanillic acid and the presence of tumour on sites as ganglia and retro-peritoneal adrenals (George *et al*., 2001). These autonomic dysfunctions as a result of catecholamine secretion occur in 37% of these cases (George *et al*., 2001). Ganglioneuroma is a slow growing tumour with so far no recorded metastatic potential and complete excision being suffice to bring about a cure. The site, size and pathological type determine the choice of surgical approach (Allison *et al*., 1989). Various approaches for removal of parapharyngeal space tumours have been described, including the transcervical approach, transparotid approach, transcervical-transpharyngeal approach, infratemporal fossa approach and combinations of the these approaches (Abemayor *et al*., 2002; Bass 1982) Chemotherapy or radiotherapy is not usually required, even for cases with partial excision (Retrosi *et al*., 2011).

In differential diagnosis a ganglioneuroma should be distinguished from the immature forms of neuroblastoma 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 tumours such as these along with carcinoid and pheochromocytoma. Immunohistochemistry is advocated, in which ganglion cells stain for neurofilament or neuron specific enolase whereas Schwann cells stain for S-100 protein (Trojanowski *et al*., 1991).

Conclusion

The pathology may not be suspected easily due to its rarity in presentation. Although very rare, ganglioneuromas must be considered in the differential diagnosis of neck masses. Ganglioneuroma of the neck, most commonly presents as an

enlarging neck mass. Surgical excision via cervical approach offers definitive therapy. And prognosis is excellent after surgical excision.

REFERENCES

Abemayor, E. and Lufkin, R. 2002. Enhancing access to the parapharyngeal space. Laryngoscope, 112:757-759

Albonico, G., Pellegrino, G., Maisano, M. and Kardon, D. E. 2001. Ganglioneuroma of parapharyngeal region. *Arch Pathol Lab Med*, 125:1217-1218

Allison, R. S., Van der Waal, I. and Snow, G. B. 1989. Parapharyngeal tumours: a review of 23 cases. *Clin Otolaryngol Allied Sci*, 14:199-203

Bass, R. M. 1982. Approaches to the diagnosis and treatment of tumors of the parapharyngeal space. Head Neck Surg, 4:281-289

Bove, K. E. and Mc Adams, A. J. 1981. Composite ganglioneuroblastoma an assessment of the significance of histologic maturation in neuroblastoma diagnosed beyond infancy. *Arch Path Lab Med*, 105:325-30.

Califano, L., Zupi, A., Mangone, G. M. and Long, F. 2001. Cervical ganglioneuroma: report of a case. *Otolaryngol Head Neck Surg*, 124:115-116.

Enzinger, F. M. and Weiss, S. W. 1995. Primitive neuroectodermal tumors and related lesions. In Soft Tissue Tumors. 3rd edition. Edited by Enzinger FM, Weiss SW. St Louis: Mosby Year-Book; pp. 929-964.

George, B., Hero, B., Harms, D., Grebe, J., Scheidhauer, K. and Berthold, F. 2001. Metabolic activity and clinical features of primary ganglioneuromas. *Cancer,* 91(10):1905-1913.

Retrosi, G., Bishay, M., Kiely, E. M., Sebire, N. J., Anderson, J., Elliott, M., Drake, D. P., Coppi, P., Eaton, S. and Pierro, A. 2011. Morbidity after ganglioneuroma excision: is surgery necessary? *Eur J Pediatr Surg,* 21:33–37

Schumaker, H. B. and Lawrence, E. A. 1939. Cervical sympathetic ganglioneuroma: Case report and review of literature. *Surgery*, 5: 572-81.

Shimada, H., Ambros, I. M. and Dehner L. P., *et al*. 1999. Terminology and morphologic criteria of neuroblastic tumours. Recommendations by the International Neuroblastoma Pathology Committee. *Cancer*, 86: 349-363.

Trojanowski, J. Q., Molenar, W. M. and Baker D. L., *et al*. 1991. Neural and neuroendocrine phenotype neuroblastoma, ganglio-neuroblastoma, ganglioneuroma and mature versus embryogenic human adrenal medullary cells. *Prog.Clin. Biol. Res,* 366: 335-341

Weber, A. L., Montandon, C. and Robson, C. D. 200*0. Neurogenic tumors of the neck. Radiol Clin North Am*, 38:1077-1090.

Weiss, S. W. and Goldblum, J. R. 2008. Enzinger and Weiss's Soft Tissue Tumors. 5th ed. St. Louis: Mosby, pp. 960–963.

****