

Ganglioneuroma at the Coccyx and the Review of Literature

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Abstract

Ganglioneuroma are complex tumours that arise in peripheral ganglia and are composed of well differentiated neurons, nerve processes, Schwann cells and enteric glial cells. They coexist with pheochromocytoma, secrete various neuropeptides producing hypertension or hypotension. These tumours are benign and they never metastasize, because of the benign nature of ganglioneuroma adjuvant chemotherapy or radiotherapy is not indicated, but regular follow-up is necessary for early diagnosis of potential local recurrence. There are prognostic markers like Tyrosine Hydroxylase and Glyceraldehyde-3-Phosphate Dehydrogenase which can be measured in blood and bone marrow.

Introduction

Ganglioneuroma is a rare benign neurogenic tumour originating from the sympathoadrenal nervous system and is considered the benign counterpart of neuroblastoma, lacking the immature neuroblastic cells.¹ They co-exist with pheochromocytoma, secrete various neuropeptide producing hypertension or hypotensive crisis during anaesthesia for operating these tumours. These tumours though benign can very rarely metastasize to regional lymph nodes or to distant sites. Ganglioneuroma usually arise in the central nervous system but its appearance in GTT mesentery is less reported. Ganglioneuroma accounts for less than 1% of all soft tissue neoplasm.² It is benign well differentiated slow growing tumour composed of ganglion cells and schwann cells. It can arise any where

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from the base of the skull to the pelvis.³ We here report a quite rare case of silent ganglioneuroma at the Coccyx.

Case Report

Male age 30 years presented with a mass at the coccyxgeal region since 2 years. Asymptomatic except for physical discomfort.

O/E-GC good

- Mass well capsulated soft measuring 4 x 3 x 4 cms.
- BP 138/90 mm Hg in supine position
- Pulse 90/min RR 20/min
- No H/O seizures H/O GTT complaints
- No H/O Tingling numbness, paraesthesia.

Investigation CBC

Hb 13.2 gm%	RBC	WBC
	4.7 mil/cumm	8700/cumm
N 67% E 02%	L 31% M 0%	
BT	CT	
3 mins 30 secs	6 mins 45 secs	
PT Test 14 secs	Control 14 secs	INR 1.0
FBS 88.7 mg%	HIV Negative	HBsAg Negative
HCV Negative	VDRL	(Prozone phenomenon Ruled out)
	Non Reactive	

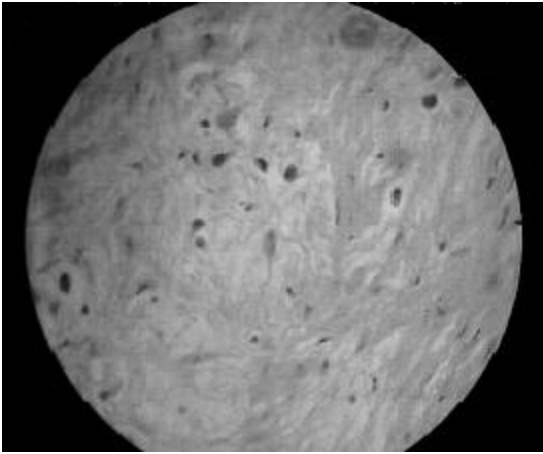


Fig. 1 :H and E stain 45x/Tumour histology of Ganglioneuroma (Schwannian Stroma-dominant) mature type

Preoperative investigation of Urine VMA was not done.

Post operative Urine VMA was 8.1 mg/24 hrs (Vinyl Mandelic acid)

Normal Range : 1.8 – 7.1 mg/24 hrs

Excision was under local anaesthesia

Histopathology finding were suggestive of ganglioneuroma, microscopically tumour histology was that of ganglioneuroma (Schwannian stroma-dominant) mature type (Fig. 1).

Discussion

Ganglioneuroma are complex tumours that arise in peripheral ganglia and are composed of well differentiated neurons, nerve processes. Schwann cells and enteric glial cells. The term Ganglioneuromatosis denotes a regional or segmental proliferation of ganglioneuromatous tissue. Both phaeochromocytomas and ganglioneuromas originate from neural crest cells.⁴ The case report showed rise of neurosecretory peptides but at very steady state, this probably may be because of slow growth, many ganglioneuromas are large when they are diagnosed and these do release excessive catecholamine. The totally asymptomatic clinical picture of the case report could be

explained by the very slow growth of tumour which can displace the surrounding anatomical structures without infiltration. History of seizures must also be correlated with noncranial ganglioneuromas.⁷

Most of noncranial Ganglioneuromas are located in the posterior mediastinum and the retroperitoneal. However, Ganglioneuroma at presacral region has been reported by Cerullo G *et al* in April 2001^{5,6} but our case report to our knowledge may be second case at the coccyx which was asymptomatic probably involving the cauda equina which could have been proved by immunohistochemistry studies for cytokeratin.⁸ Assuming preoperative urinary VMA and S. catecholamine level to be normal but post operative the levels were high which could have caused hypertensive crisis¹ but patient was operated under local anaesthesia. The levels were high because of manipulation during excision. Ganglioneuromas should be kept as the D/D for any solid tumour mass in the mediastinum, retroperitoneal, GIT, goitre swelling as screened in the literature and one should study neuropeptide levels preoperatively which would be cost effective rather than doing immunohistochemistry staining.^{9,10}

Ganglioneuromas diagnosed individually, history of exposure especially syphilis to be considered as important parameters in adult cases since *Treponema palladium* may induce a tetragenic and oncogenic effect.¹¹

Ganglioneuroma mimicking ovarian tumours have been reported which could have been diagnosed by preoperative neuropeptide estimation, alpha foeto protein, beta hCG^{12,13} Ganglioneuroma are relatively rare and difficult to distinguish from other tumours due to lack of image findings specific for ganglioneuroma.³

Ganglioneuroma can be surgically

dissected with favourable prognosis but preoperative D/D is sometimes difficult which could be overcome by doing preoperative estimation of neuropeptides.¹⁴

Because of the benign nature of Ganglioneuroma and since they rarely metastasize.⁵ Adjuvant chemotherapy or radiotherapy is not indicated but regular follow up is necessary for early diagnosis of potential local recurrence. For this reason we should maintain a therapeutic criterion for carrying out a biopsy, as well as a follow up and control if the histological studies do not show up any kind of malignancy.¹⁵ Further studies at molecular level are by reverse transcription PCR using a new real time detection method. We can measure 2 genes viz tyrosine hydroxylase (TH) as the target gene and glyceraldehydes 3 phosphate dehydrogenase as a reference gene in blood and bone marrow as prognostic marker.

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