■ *Case Report*

Retroperitoneal ganglioneuroma

S Karki[1], AK Sinha[1], P Upadhyaya[1], M Agarwal[1], A Pradhan[1], P Paudyal[1], SU Kafle[1], G Kattel[1], RK Agarwal[2]
[1]Department of Pathology and [2]Surgery
B.P.Koirala Institute of Health Sciences, Dharan, Nepal

Abstract

Ganglioneuromas are fully differentiated benign tumors of the sympathetic nervous system arising from the neuroectodermal cells. Its reported incidence is one per million populations. We report an incidentally detected, histopathologically proven ganglioneuroma of the retroperitoneum in a 39 year old lady with the preoperative diagnosis of adrenal tumor.

Keywords: ganglioneuroma, retroperitoneum, neuroectodermal cells

Background

Ganglioneuromas are tumors of the sympathetic nervous system that arise from the neuroectodermal cells.[1] Neuroblastoma, ganglioneuroblastomas and ganglioneuromas are tumours within the spectrum of its disease continuum. Ganglioneuromas are fully differentiated benign tumours. The reported incidence is one per million population.[1] The most common localization is the posterior mediastinum followed by the retroperitoneal space.[1,2] Among the primary retroperitoneal tumors, they constitute only a small percentage of 0.72 to 1.6.[1,3] Although catecholamine synthesis is an almost constant feature of all the neurogenic tumors, ganglioneuromas rarely lead to symptoms.

Case report

We report an incidental detection of retroperitoneal tumour in a 39 year old lady who presented to the medical and surgical OPD with the complaint of episodic epigastric and right lumbar pain since a period of three years.

Her laboratory investigations for complete blood count, serum electrolytes and urine were normal. Ultrasonography and CT scan of the abdomen incidentally showed a heterogenous mass in the region of the right adrenal gland extending below the inferior vena cava.

A clinicoradiological diagnosis of adrenal tumor was made. The patient underwent exploratory laparotomy for removal of the tumour and the material was submitted for histopathologic examination. Grossly the tumor measured 8.5x7x3.2 cm. It was capsulated, firm in consistency with a homogenous, solid, grayish white cut surface and gritty sensation (Figure 1 & 2). Microscopically, the tumor was that of a ganglioneuroma (schwannian stroma-dominant) mature type with focal areas of calcification (Figure 3 & 4).

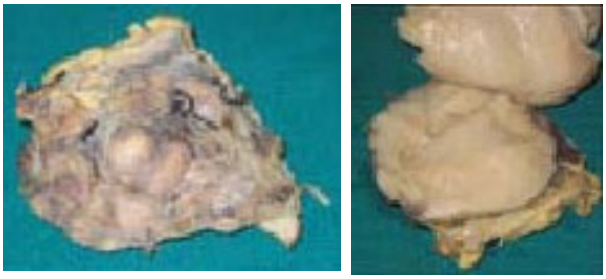

Figure 1. Ganglioneuroma. Excised capsulated tumour at gross appearance.

Address for correspondence
Dr Smriti Karki
Associate Professor, Department of Pathology, BPKIHS
Email: dr_smritikarki@yahoo.com

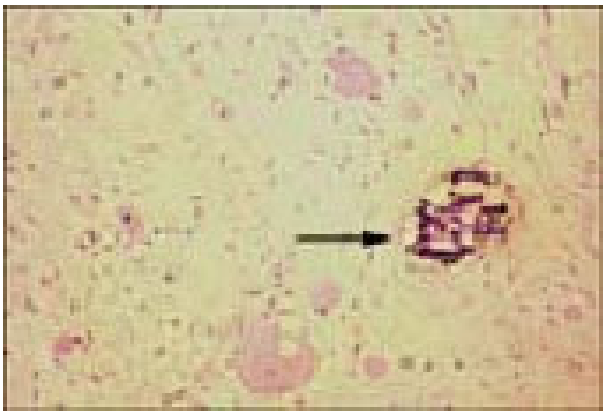

Figure 2. Grossly a circumscribed tumour with homogenous whitish yellow cut surface.

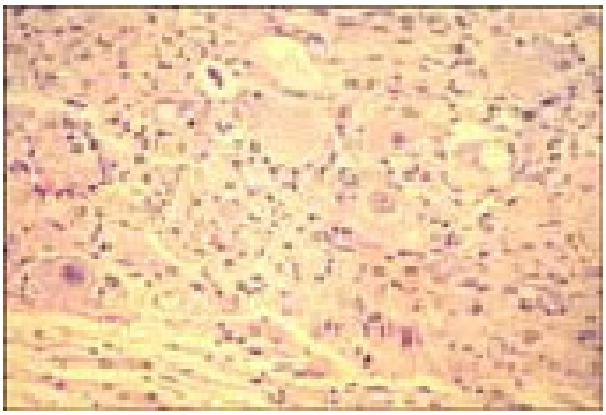

Figure 3. Photomicrograph showing presence of schwannian stroma, large calcification and ganglion cells. 200X; H&E.

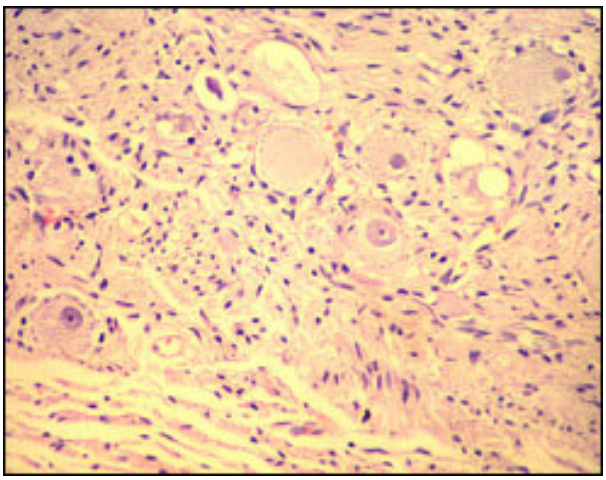

Figure 4. Ganglioneuroma showing presence of ganglion cells and schwannian stroma 400X; H&E.

Discussion

Ganglioneuromas usually appear in children, while retroperitoneal ganglioneuromas appear more often in adults. The clinical symptoms of GNs are non-specific, mostly hormonally silent.[4,5] Majority of cases are detected incidentally since they are usually asymptomatic.[6]

Tevfik reported incidental detection of retroperitoneal GN in 12 year old boy.[7] These tumours may come to attention due to compression of their neighboring structures. Symptoms of autonomic dysfunction may be caused by paravertebral ganglioneuromas compressing the autonomic fibers of the lumbosacral plexus. Symptoms like diarrhea, sweating and hypertension may be related to release of peptides like vasoactive intestinal peptide (VIP), somatostatin and Neuropeptide Y.[8]

The trend of utilizing radiology as a screening technique is increasing the detection of retroperitoneal tumours. However the preoperative diagnosis of Ganglioneuroma is difficult as imaging of these tumours is non specific and nondiagnostic.[9] On reviewing literatures most of such tumours are preoperatively diagnosed as retroperitoneal tumour presenting as renal mass or adrenal mass as is in the index case and also uniquely as pelvic tumour with retroperitoneal extension.[1,10,11]

Cytologys an investigative tool though reported to be useful in the preoperative diagnosis of ganglioneuromas has its limitation. This is so when there are less well differentiated areas within the tumour which demands surgical exploration and histopathological examination. Hence biopsy is not only of diagnostic value but also of prognostic significance.

Grossly, they are large, encapsulated masses of firm consistency with an homogenous, solid, grayish white cut surface. Areas with different colour or consistency should be sampled for microscopic examination with the suspicion of less differentiated foci.[12]

The International Neuroblastoma Pathology Committee (INPC) has divided Ganglioneuromas into two subtypes :*GN maturing* and *GN mature*. The morphologic classification is prognostically

significant. Ganglioneuroblastoma and Neuroblastoma are less mature and more aggressive as these tumours have a high neuroblast content.

Microscopically the overall appearance of a Ganglioneuroma is that of a neurofibroma (hence, its designation Schwannian stroma dominant tumour). If every single ganglion cell is mature the designation mature subtype. If there is maturing/ minor component of scattered collections of differentiating neuroblast, the tumour is designated maturing subtype.[14]

Exceptionally component of the ganglioneruma may show features of malignant nerve sheath tumour.[13] GNs are slow growing benign tumours which can be surgically dissected with favourable prognosis. Local recurrence has been reported, so periodic radiologic surveillance is perfromed after resection.

Conclusion

The widespread use of radiological investigations has increased the detection of retroperitoneal tumours. However, the pre operative diagnosis of Ganglioneuroma is difficult. Therefore ganglioneuroma is always a histopathological diagnosis.

References

1. Lamichhane N, Dhakal HP. Ganglioneuroma of pelvis- an unique presentation in a young man. Nepal Med Coll J. 2006 Dec; 8 (4):288-91
2 . T S, Michalopoulos N, Karayannopoulou G, Kesisoglou I ,Valentini T , Raptou G; Papavramidis S T. Retroperitoneal Ganglioneuroma in an Adult Patient: A Case Report and Literature Review of the Last Decade. Southern Medical Journal. 2009 October; 102 (10):1065-1067.
3. Moriwaki Y, Miyaka M, Yamamoto T *et al*. Retroperitoneal ganglioneuroma: a case report and review of the Japanese literature. *Intern Med* 1992; 31: 82-5.
4. Hayes FA, Green AA, Rao BN. Clinical manifestations of ganglioneuroma. *Cancer* 1989; 63: 1211-4.
5. Shimada H, Ambros IM, Dehner LP, Hata J, Joshi VV, Roald B. Terminology and morphologic criteria of neuroblastic tumors: recommendations by the International Neuroblastoma Pathology Committee. Cancer. 1999;86:349-63.
6. Sucandy I, Akmal YM, Sheldon DG. Ganglioneuroma of the adrenal gland and retroperitoneum: A case report. North Am J Med Sci 2011; 3: 336-338.
7. Tevfik Aktoz, Mustafa Kaplan, Ufuk Usta, Ýrfan Hüseyin Atakan, Osman Ýnci. Retroperitoneal ganglioneuroma. Trakya Univ Tip Fak Derg 2009;26(2):163-165
8. Lonergan GJ, Schwab CM, Suarez ES, Carlson CL. Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma: radiologic-pathologic correlation. *Radiographics* 2002;22:911-934.
9. Bjellerup P, Theodorsson E, Kogner P. Somatostatin and vasoactive intestinal peptide in neuroblastoma and ganglioneuroma: chromatographic characterization and release during surgery. *Eur J Cancer* 1995;31A:481-485.
10. Yuji H, Osamu Y and Koichi H. Retroperitoneal ganglioneuroma. Nishinihon Journal of Urology 2003; 6(6):376-379.
11. Papavramidis T S, Michalopoulos N, Karayannopoulou G, Kesisoglou I ,Valentini T, Raptou G; Papavramidis S T. Retroperitoneal Ganglioneuroma in an Adult Patient: A Case Report and Literature Review of the Last Decade. Southern Medical Journal. 2009 October; 102 (10):1065-1067.
12. Rosai J. Chap 16: Adrenal gland and other paraganglia. In: Ackermans surgical pathology, vol 1, 8th edn. Mosby, St. Louis; 1996. pp1015-58.
13. Koss, Brown LP, Randin R, Shibata D, Chandrasoma P Malignant peripheral nerve sheath tumour arising in an adult male homosexual. Cancer. 1986;57:2022-2025.
14. Ronald A, De Lellis. The adrenal glands. In: Sternberg's Diagnostic Surgical Pathology. Shamlas Mangray, Darryl Carter, Mills. Stacey E editors. Ganglioneuroma (Schwannian Stroma dominant neuroblastic tumour) Ed 4th Lippincott, Philadelphia. 2006;p.650-651.