International Surgery Journal
Jain AA et al. Int Surg J. 2020 Aug;7(8):2737-2740
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20203266

Case Report

Ganglioneuroma mimicking adrenal tumor

Abhijeet A. Jain*, Girish D. Bakhshi, Jessika Shah, Dinesh Pawar, Aditya B. Marathe, Avinash Gonnade, Abhay Kedare, Parth B. Gada

Department of Surgery, Grant Government Medical College and Sir J.J. Group of Hospitals, Mumbai, Maharashtra, India

Received: 09 January 2020
Revised: 11 July 2020
Accepted: 15 July 2020

***Correspondence:**
Dr. Abhijeet A. Jain,
E-mail: jain.abhijeet112@gmail.com

ABSTRACT

Ganglioneuromas arise from neural crest sympathogonia and are rare benign neurogenic tumors. The most common affected sites are posterior mediastinum and the retroperitoneum. They rarely affect adrenal glands. Ganglioneuromas often present as solitary, painless and slow growing mass and are benign in nature. These tumors are closely related to major vessels. Hence, surgical management of retroperitoneal pathologies may require multiorgan resection in order to achieve complete surgical resection while preservation of surrounding organs especially in case of benign tumors. We present a case report of a 21 year old male with a 12×10×10 cm size right sided retroperitoneal ganglioneuroma which on computed tomography (CT) mimicked adrenal tumor crossing the midline and abutting the aorta and splaying the inferior venal cava and renal vein. Present paper is an attempt to review the various surgical options available while dealing with these benign retroperitoneal tumors which are related closely to retroperitoneal organs and major vessels.

Keywords: Retroperitoneal tumor, Adrenal gland, Inferior vena cava, Surgery

INTRODUCTION

Ganglioneuromas are benign slow growing tumors. These are commonly seen in retroperitoneum and mediastinum the retroperitoneum harbors various vital structures like great vessels, kidneys, adrenals, ureters, ascending and descending colons and the ureters. This requires extreme caution and expertise while dealing with tumors arising in retroperitoneum. Management of such tumors often requires multi organ resection and complex reconstruction for adequate resection.[1] Careful dissection and preservation of these structures is paramount, especially in case of a benign mass. A case of 21 years old male presenting with right sided retroperitoneal tumor of size 12×10×10 cms compressing the inferior vena cava (IVC), abutting aorta, right renal vein and mimicking tumor of adrenal gland is presented. Imaging may not be helpful sometimes to differentiate ganglioneuroma from adrenal gland tumor. These tumors require meticulous dissection owing to large size and close relation with major vessels, thus preserving the structures and avoiding the need for complex reconstructions. Surgical excision is treatment of choice for large tumors. A case report with review of literature and clinical approach is presented.

CASE REPORT

A 21 years old male, presented with pain in abdomen not interfering with daily activities since 6 months. He had history of trivial trauma 1 year back. He had no associated bladder or bowel complaints. Patient had no history of sweating, palpitations, syncope, headache. Patient had no previous history of surgery or radiation exposure. Clinical examination revealed normal vital parameters with soft abdomen and no palpable or ballotable lump. His ultrasound of the abdomen was

suggestive of a cystic lesion measuring 12×10×4.7 cm in size in the right adrenal gland. In view of trauma initial differential was hematoma or adrenal cyst. Contrast enhanced CT scan with Ultrasound correlation revealing a hypodense non enhancing lesion with peripheral calcification in the vicinity of right adrenal gland measuring 12×10.8×10.7 cm not separate from right adrenal gland abutting the liver with no invasion. There was loss of fat plane between tumor and inferior vena cava and dilatation of right renal artery due to compression by the tumor. Tumor was observed to cross the midline posterior to inferior vena cava and abutting the aorta (Figure 1). Left adrenal gland was normal.

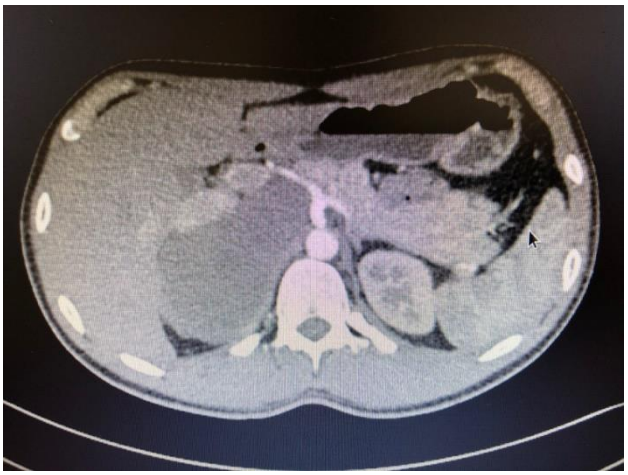

Figure 1: CT scan showing right ganglioneuroma pushing inferior venacava and aorta.

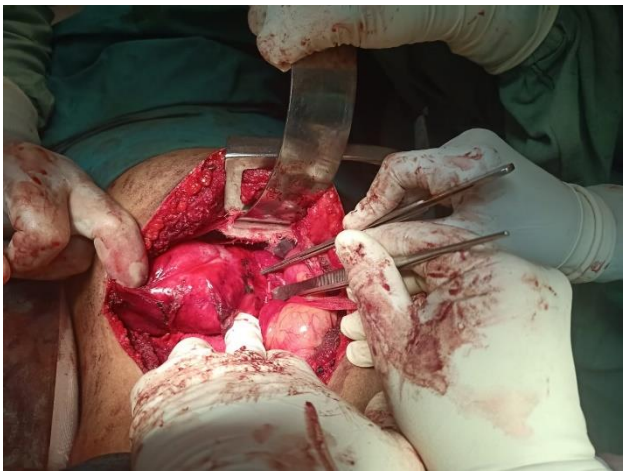

Figure 2: Intraoperative picture showing tumor extending below inferior venacava and compressing rt renal vein.

Patient underwent a CT guided biopsy of the lesion with histopathology report suggestive of smooth muscle tissues not representative of the mass. Patient due to personal reasons wanted discharge and was told to follow up after 6 weeks. Repeat contrast enhanced CT scan after 6 weeks was suggestive of no significant interval change. His serum and urine catecholamine and their metabolite levels were unremarkable and ruled out functional tumor of adrenal gland.

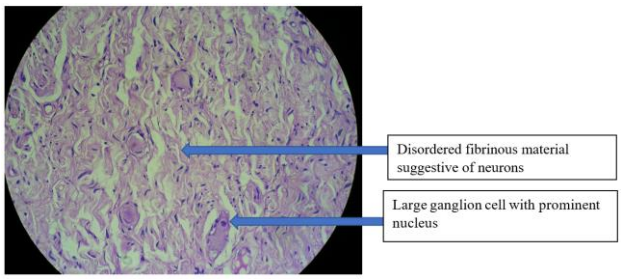

Figure 3: Histopathology slide showing large ganglion cells with prominent nucleus magnification: 40x hematoxylin and eosin stain.

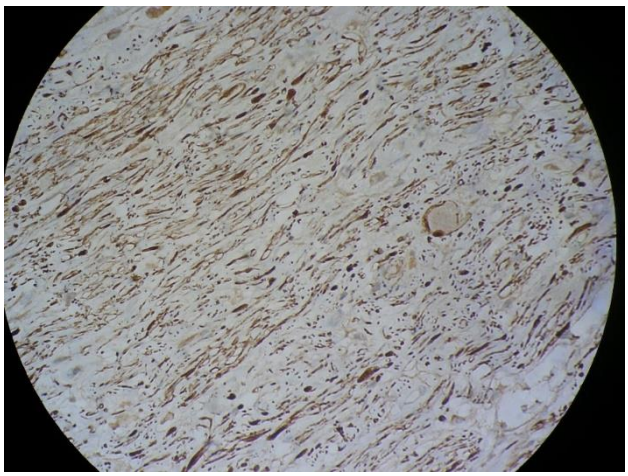

Figure 4. Immunohistochemistry showing synaptophysin positive.

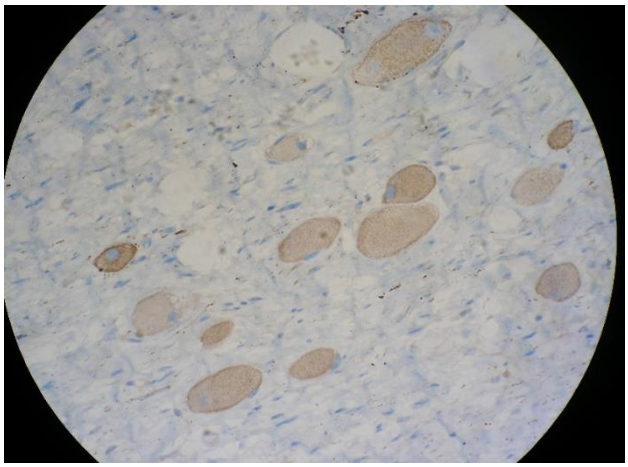

Figure 5. Immunohistochemistry showing S-100 positive.

Decision was taken to post the patient for surgery as the mass was abutting the major vessels and the histopathology report through minimally invasive method was inconclusive. Open surgical excision was planned in view of size of the tumor and compression of the inferior

vena cava. Supra coastal incision was taken and twelfth rib was excised. Tumor was seen to be abutting the liver superiorly and crossing the midline abutting the aorta and pushing the inferior vena cava anteriorly. Inferior vena cava was hooked to facilitate the excision (Figure 2) Adrenal was seen separate from the lesion and appeared to be splayed by it. Medial branches to the tumor from the aorta were lighted and tumor was dissected off the inferior vena cava and was delivered in mass.

On gross examination, the tumor measured 11×9×10 cm size with cut section showing myxoid whitish solid areas. On histopathological examination there were large ganglion cells with prominent nuclei with eosinophilic granular bodies and disordered fibrinous materials which was suggestive of ganglioneuroma (Figure 3). Immuno-histochemistry was positive for synaptophysin (Figure 4) and S-100 (Figure 5) thereby confirming the diagnosis. Postoperative course was uneventful. Follow up of 9 months has shown patient to be disease and symptom free.

DISCUSSION

Ganglioneuroma is a rare tumor arising from neural crest which is composed of gangliocytes and mature stroma. Most common sites of presentation are posterior mediastinum, retroperitoneum and head and neck region. 15 to 30% occur in adrenal glands.[2]

The presented case was a ganglioneuroma in the retroperitoneum in the vicinity of the adrenal gland which presented a diagnostic challenge as the tumor could not be seen separately from the adrenal gland by radiological investigations.

Prevalence of ganglioneuroma in a population is 1 per million. As a primary retroperitoneal tumor ganglio-neuroma constitutes 0.72 to 1.6%.[3] In more than 40% of patients tumor is poorly symptomatic or completely asymptomatic and may grow slowly without producing symptoms. Some tumors exhibit hormonal activity and present a broad range of symptoms depending upon the hormones. They are usually present in younger age group and are discovered incidentally or due to non specific symptoms caused by their mass effect on their adjacent organs.[4]

The patient though asymptomatic had grossly dilated inferior vena cava and renal veins and was observed to be abutting the major vessels and compressing them hence warranting excision.

There are several features suggesting a ganglioneuroma in case of an adrenal tumour like lack of hormonal secretion, evidence of calcification, no vessel involvement.[5] High levels of urinary catecholamines help to differentiate between ganglioneuroma and neuro-blastoma.[6] The presented case was thoroughly evaluated for serum and urine catecholamine levels along with meticulous history taking to rule out functional tumor.

Ganglioneuromas have a characterstic low attenuation on unenhanced CT scans.[7] Although high index of suspicion can aid in diagnosis based on radiological findings, surgery is recommended for all incidentalomas larger than 5 cm as only tissue diagnosis can distinguish between various adrenal pathologies.[8]

Complete surgical excision reaching negative tissue margin results in excellent outcome and is the treatment of choice in malignant cases.[9] The surgical resection of these tumors can be undertaken via laparoscopic or open technique. Laparoscopic surgery is increasingly becoming common for retroperitoneal pathologies due to increasing expertise and advanced energy sources and instruments. In present case open surgery was conducted owing to proximity of the tumor to major vessels as well as large size of the tumor.

Alimoglu et al described en bloc resection of 50 mm retroperitoneal tumor with careful dissection from celiac trunk, segment 1 of liver, pancreas and left gastric and hepatic arteries.[10] Abraham et al have reported excision of 13×9.5×6 cm and 17×11×7.5 cm tumors.[11] These suggest that laparoscopic excision is possible even of large tumors with shorter hospital stay and fewer complications.

Open surgeries have been preferred while operating in proximity to great vessels where laparoscopy may face limitations. Vasiliadis et al reported an extra adrenal ganglioneuroma involving intrahepatic IVC, superior mesenteric artery and celiac axis which was excised via an upper midline laparotomy.[12]

Some novel techniques like fractionated resection by Wan et al and hanging method by Oue et al have also been described.[13,14] Major vascular resections and prosthetic replacements have also been reported by Fueglistaler et al.[15]

No matter what the mode of surgery is chosen, there should be a low threshold for conversion to open and oncological principles must be followed for desirable outcomes.

Microscopically, ganglioneuromas consists of spindle cell tumour composed of neuritic processes, schwann cells and perineural cells and show numerous ganglion cells.[16] Immunohistochemistry shows schwann cell/ stroma positive for S100, synaptophysin and ganglion cells positive for S100, synaptophysin and chromogranin A.[17]

The present case was diagnosed as ganglioneuroma based on the classical histological and immunohistochemistry features.

CONCLUSION

Present case shows that ganglioneuromas may mimic primary adrenal tumor. Radiological imaging helps in planning surgical excision. Complete surgical resection using meticulous and novel surgical techniques with adherence to oncological principle helps in preserving the surrounding vital structures. Histology complimented by immunohistochemistry helps in establishing final diagnosis. With increasing expertise, this may be achieved laparoscopically as well but each case must be individualized on case to case basis without compromising on safety and oncological principles.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Strauss DC, Hayes AJ, Thomas JM. Retroperitoneal tumors: Review of management. Ann Royal Coll Surg England. 2011;93:275-80.
2. Felix EL, Wood DK, DasGupta TK. Tumours of the retroperitoneum. Curr Probel Cancer. 1981:6:1-47.
3. Shetty PK, Balaiah K, Gnana Prakash S, Shetty PK. Ganglioneuroma Always a histopathological diagnosis. OJHAS. 2010;9:19.
4. Hayes FA, Green AA, Rao BN. Clinical manifestation of ganglioneuroma. Cancer. 1989;63:1211-4.
5. Maweja S, Materne R, Detrembleur N, de Leval L, Defechereux T, Meurisse M, et al. Adrenal ganglioneuroma. Am J Surg. 2007;194:683-4.
6. Georger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic acitivity and clinical features of primary ganglioneuroma. Cancer. 2001;91:1905-13.
7. Ichikawa T, Ohomoto K, Arkari T, Fujimoto H, Nemoto K, Nanbu A, et al, Ganglioneuroma: computed tomography and magnetic resonance features. Br J Radiol. 1997;69:114-21.
8. Yeh MW, Livhits MJ, Duh QY. The Adrenal Glands. Sabiston Textbook of Surgery, First South Asian Edition. 2017.
9. Linos D, Tsirlis T, Kapralou A, Kiriakopoulos A, Tsakayannis D, Papaioannou D. Adrenal ganglioneuromas: incidentalomas with misleading clinical and imaging features. Surgery. 2011;149:99-105.
10. Alimoglu O, Caliskan M, Acar A, Hasbahceci M, Canbak T, Bas G. Laparoscopic excision of retroperitoneal ganglioneuroma. J Society Laparo-endoscopic Surgeons. 2012;16:668-670.
11. Abraham GP, Siddaiah AT, Das K, Krishnamohan R, George DP, Abraham JJ, et al. Laparoscopic extirpation of giant adrenal ganglioneuroma. J Minimal Access Surg. 2014;10:45-7.
12. Vasiliadis K, Papavasiliou C, Fachiridis D. Retroperitoneal extra adrenal ganglioneuroma involving the infrahepatic inferior venal cava, celiac axis and superior mesenteric artery: A case report. Int J Surg Case Reports. 2012;3:541-3.
13. Wan Z, Yin T, Chen H, Li D. Surgical treatment of retroperitoneal benign tumor surrounding important blood vessels by fractionated resection: A case report and review of literature. Oncology Letters. 2016;11:3259-64.
14. Oue T, Yoneda A, Sasaki T, Tani G, Fukuzawa M. Total laparoscopic exision of retroperitoneal ganglioneuroma using hanging method and vessel sealing device. J Laparoscopic Adv Surg Tech.2008;18:779-82.
15. Fueglistaler P, Gurke L, Stierli P, Obeid T, Koella C, Oerti D, et al. Major vascular resection and prosthetic placement for retroperitoneal tumors. World J Surg. 2006;30:1344-9.
16. Genta S, Rainero I, Luda E, Pinessi L. Autonomic dysfunction associated with multiple pelvic ganglioneuromas. Acta Neurol Scand. 2001;104(1):54-6.
17. Dabbs DJ. Diagnostic immunohistochemistry, Elsevier Publications, 3rd edition., 2010.

Cite this article as: Jain AA, Bakhshi GD, Shah J, Pawar D, Marathe AB, Gonnade A, et al. Ganglioneuroma mimicking adrenal tumor. Int Surg J 2020;7:2737-40.