ORIGINAL RESEARCH PAPER

Surgery

KEY WORDS:

LESSER OF TWO EVILS – A CASE SERIES ON GANGLIONEUROMAS

Dr Mohanavel MS MCh - Associate professor of paediatric surgery – Villupuram medical college

Dr V Rohit Gopinath* MS General Surgery, Mch paediatric surgery, Asst professor Department of paediatric surgery, ICH *Corresponding Author

Prof J Muthukumaran MS General surgery, MCh paediatric surgery, HOD Department of paediatric surgery ICH and HC

ABSTRACT

Ganglioneuroma is the benign counterpart in the spectrum of peripheral neuroblastic tumours. Here I am presenting 4 cases of this rare tumour.1st is a 2 year old female child who came with complaints of fever and abdominal distension of 2 week duration. On Examination of the abdomen there was no mass palpable. USG abdomen done showed a mass arising from the right adrenal gland. CECT abdomen was done which showed a heterogenous mass arising from the right adrenal gland abutting the IVC . Initially a laparotomy and biopsy was done. It was reported as ganglioneuroma .Oncology opinion was got and follow up was done monthly for 3 months . Relaparotomy was done and excision of the tumour was done.2nd child is a 7 year old female child who was brought with complaints of abdominal pain on and off and fever of 2 months duration. On examination of the abdomen there was no mass palpable .USG abdomen done showed an enlarged left para aortic .CECT abdomen done showed an enlarged paraaortic node with necrotic changes .She underwent a laparotomy and excision of the paraaortic mass. HPE was reported as ganglioneuroma. The other 2 cases have been summarised.

INTRODUCTION

Ganglioneuroma is a rare benign tumour arising from neural crest sympathogonia which are undifferentiated cells of the sympathetic nervous system. These do not contain any immature elements. These tumours most commonly occur in the abdomen , however they can be seen in any area where sympathetic nervous tissue is seen. Other areas include adrenal gland , retroperitoneum , posterior mediastinum, head and neck. It is interesting to note that, thoracic tumors have been found to be larger than non-thoracic ones at the time of diagnosis. Its occurrence is equal in both males and females. A familial predisposition and an association with Turner syndrome and multiple endocrine neoplasia II have a been documented. It is important to note that Adrenal GNs are hormonally silent and as a result can be asymptomatic; even when the lesion is of a large size. However, It has been reported that up to 30% of patients with GNs may have elevated plasma and urinary catecholamine levels without showing any symptoms of catecholamine excess. Additionally, it has been documented that ganglion cells can secrete vasoactive intestinal peptide (VIP), while pluripotent precursor cells sometimes produce steroid hormones which include cortisol and testosterone.

Here we are documenting four cases of this uncommon benign variant of a common malignancy of childhood.

CASE 1

The first child was a 5year old female who was brought by her mother with complaints of loose stools and abdominal pain of 3 months duration. The child did not have any other gastrointestinal symptoms.She had been evaluated outside where in she was found to have mass arising from the right adrenal gland ? neuroblastoma which was deemed to be inoperable and only an open biopsy was done in 2017 . She was subsequently referred to our institute for further management .On examination she was pale and had a normal blood pressure for age. She was not dehydrated . On examining her abdomen there was mild distension noted but there was no obvious mass palpable. She had an abdominal scar from the previous open biopsy done. She had a haemoglobin level of 7.6gm%. Serum electrolyte levels were found to be normal. 24 hour VMA level was normal. Skeletal survey done showed no skeletal metastasis . Bone marrow aspiration done showed no marrow involvement. USG abdomen done showed a mass arising from ? Right adrenal gland of size 7.2 x 5.4x 5.5 cm. The opposite adrenal gland was found to be normal. There was no free fluid abdomen . CECT abdomen done showed a heterogeneously enhancing mass arising from the right adrenal gland of size 5.4 x 5 x 4.5 cm. There was no encasement of major vessels or infiltration of liver . Oncology opinion was obtained and it was suggested that the lesion be excised. The child was taken up for a laparotomy with right adrenalectomy . Intraoperatively the mass was found to be closely abutting the IVC and was dissected from the same. Complete excision of the mass was done. Post operatice period was uneventful and the child was discharged on post operative day 5. HPE was reported as ganglioneuroma . The child is on regular follow-up and is asymptomatic.

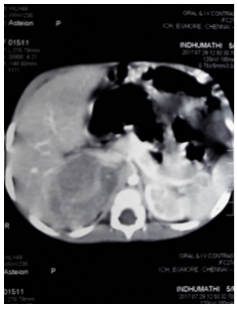

Fig 1

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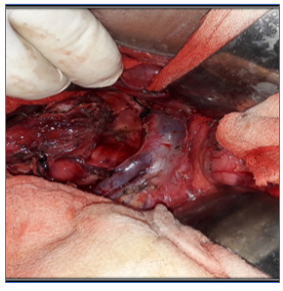

Fig 2

Case 2

The 2nd child was a 7 year old female who was brought by her mother with history of abdominal pain , low grade fever of 2 months duration. She also had history of decrease in appetite with loss of weight . She was found to have no other comorbidities or contact with TB . On examination she was found to be pale with no palpable abdominal mass. Respiratory system examination was also found to be normal .There were no peripheral palpable lymph nodes. She was found to have a haemoglobin level of 9.0gm% with normal electrolyte levels.

ESR level was found to be normal and Mantoux was negative. Chest x ray done showed no abnormality. USG abdomen done showed a well defined oval heterogenous mass of size 2.8 x 2.1 cm in the left para aortic region along with multiple mesenteric nodes. CECT abdomen done showed a non enhancing mass of size 2.5x 3 cm abutting the left common iliac artery. With a preoperative differential diagnosis of lymphoma or Tuberculosis. She was taken up for a laparotomy and excision of the mass. Intra operatively she was found to have a 2.5 x 3 cm mass of rubbery consistency in close proximity to the left iliac artery. The was excised in toto. HPE was reported as ganglioneuroma maturing subtype. Post operative period was uneventful and was discharged

on post operative day 5. Subsequently oncology opinion was got and it was suggested to keep the child on follow up .The child is now asymptomatic and on regular follow up.

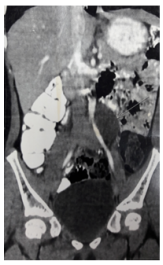

Fig 3

[Text]

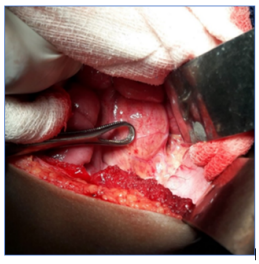

Fig 4

Other cases

Sno	Age	Complaints	Examination	Investigations	Procedure	Post op	HPE	now
3	3yrs 4 months	Abdominal pain x weeks	Right lumbar mass	24 hr VMA – negative CECT abd – 6x 5 cm mass – R Adrenal gland	Laparotomy and right Adrenalectomy done	uneventful	HPE - F/S/O Ganglioneuroma	Follow up
4	6 years 2months	Fever and Abdominal pain x1 month	No abnormalities	24 Hr VMA negative CECT Abdomen – 4 x 3 cm mass L Adrenal gland	Laparotomy and left adrenalectomy Done	Uneventful	HPE – F/S/O Ganglioneuroma	Follow up

DISCUSSION

Ganglioneuromas (GNs) are rare, differentiated tumors which arise from neural crest cells[1). They are most commonly arise from the adrenal gland . retroperitoneal space (32%-52%) or the posterior mediastinum (39%-43%). Less commonly, GNs can be seen in the cervical region (8%-9%) as well [2,3]. It has been documented that, thoracic tumors are larger than non-thoracic ones at the time of diagnosis[4]. Adrenal GNs occur mostly in the fourth and fifth decades of life, whereas GNs of the retroperitoneum and posterior mediastinum are usually seen in children and young adults. A familial predisposition as well as an association with Turner syndrome and multiple endocrine neoplasia II have also been documented. These lesions are commonly asymptomatic and can remain asymptomatic even though an increase in catecholamines production is present ,in upto 30% cases. Secretion of vasoactive intestinal peptide (VIP) and steroid hormones has also been documented.

GNs are usually discovered incidentally due to the widespread use of computed tomography and MRI imaging techniques [2]. It is to be noted that GNs account for approximately 0.3%-2% of all adrenal incidentalomas [2- 4]. In most cases, ultrasonography reveals a well-circumscribed, homogenous, hypo-echogenic lesion [5]. CT abdomen done usually shows a well-defined, lobular-shaped, solid, encapsulated mass. These tumors can range from iso-attenuating to hypo-attenuating lesions compared to muscle signals [5]. The mass can surround major blood vessels without compression or occlusion [6]. Fine, punctate calcifications are seen in 20% to 69% of cases and are considered to be diagnostic of GNs [5]. On MRI , T1-weighted images tend to have homogeneously low or intermediate signal, whereas T2-weighted images have heterogeneously intermediate or high signal [7]. The latter is said to be due to the presence of the myxoid matrix along with a relatively low number of ganglion cells [8]. Gadolinium administration can result in delayed enhancement of the lesion.

GNs are histologically benign lesions and can be divided into two main types [4]. "mature type" GNs comprise of mature Schwann cells, ganglion cells and perineural cells within a fibrous stroma whilst completely lacking neuroblasts and mitotic figures . "maturing type" GNs consist of similar cellular populations with miscellaneous maturation degrees, ranging from fully mature cells to neuroblasts.

GNs are positive for specific markers such as S-100, vimentin, synaptophysin and neuron-specific enolase.

As far as management is concerned, it is to be kept in mind that when dealing with adrenal incidentalomas of more than 6 cms , there is a 25% chance of the lesion being an adrenocortical carcinoma. Geoerger et al[4] documented local lymph node involvement in two GN patients and one case of distant metastasis to soft tissues in their 49-patient case series. Nonetheless, malignant GNs remain extremely rare [2]. Surgery is the gold standard for the treatment of GNs[4]. Even though, laparoscopic excision is usually the procedure of choice factors like hormonal activity, tumor location, and proximity to adjacent structures need to be taken into account when deciding on the best approach to operate on these rare tumors . Since GNs rarely metastasize or recur wide excision procedures are not usually required . Postoperatively, there is no need for adjuvant therapy in these patients and their prognosis is excellent [4].

CONCLUSION

GNs are uncommon, differentiated tumors which originate from neural crest cells. These lesions are usually discovered incidentally and tend to be hormonally inactive. Surgical excision is the gold standard for the treatment of GNs but preoperative diagnosis of this variant is extremely challenging. Histopathologic examination is absolutely necessary to confirm this rare diagnosis. Postoperatively, there is no need for any adjuvant chemotherapy and the overall prognosis of these patients is excellent.

REFERENCES

1. Joshi VV. Peripheral neuroblastic tumors: pathologic classification based on recommendations of international neuroblastoma pathology committee (Modification of shimada classification) Pediatr Dev Pathol. 2000;3:184–199.
2. Rha SE, Byun JY, Jung SE, Chun HJ, Lee HG, Lee JM. Neurogenic tumors in the abdomen: tumor types and imaging characteristics. Radiographics. 2003;23:29–43.
3. Zografos GN, Kothonidis K, Ageli C, Kopanakis N, Dimitriou K, Papaliodi E, Kaltsas G, Pagoni M, Papastratis G. Laparoscopic resection of large adrenal ganglioneuroma. JSLS. 2007;11:487–492.
4. Geoerger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. Cancer. 2001;91:1905–1913.
5. Qing Y, Bin X, Jian W, Li G, Linhui W, Bing L, Huiqing W, Yinghao S. Adrenal ganglioneuromas: a 10-year experience in a Chinese population. Surgery. 2010;147:854–860
6. 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.
7. Li L, Shao J, Gu J, Wang X, Qu L. Adrenal ganglioneuromas: experience from a retrospective study in a Chinese population. Urol J. 2014;11:1485–1490.
8. Allende DS, Hansel DE, MacLennan GT. Ganglioneuroma of the adrenal gland. J Urol. 2009;182:714–715.