NEUROENDOCRINE TUMOR PRESENTING AS PERITONEAL MASS- DIAGNOSTIC DILEMMA

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ABSTRACT

Neuroendocrine tumours (NET) are found to be aggressive and extremely rare tumour. Its annual incidence is approximately 2.5-5.0 per 100,000. NET which are arise from cells of the nervous and endocrine systems. Majority of NET are seen within the GIT(55%) and respiratory system(30%). Most common site are mainly 45% of small intestine,followed by 20% of rectum , 17% of appendix, 11% of colon and then 7% of stomach. Cancer of this type is highly aggressive, and can occur in younger patients.MRI – the best method to achieve because it provides precise information and clear images regarding their location, size and complexity of mass. Small cell NET is associated with thier early metastases to lymph nodes and liver and poor prognosis. Treatment for localized small cell NE carcinoma are done by mainly postoperative chemotherapy or radiation therapy with surgical excision. This type of tumor are characterized or identified by their aggressive behavior, natural history and often have widespread metastatic nature on presentation.

KEYWORDS: Neuroendocrine tumours, Small intestine, Carcinoid, Necrosis, Metastasis.

INTRODUCTION

Neuroendocrine tumours (NET) are found to be aggressive and extremely rare tumour. Its annual incidence is approximately 2.5-5.0 per 100,000(1). NET are mainly arises from cells of the nervous and endocrine systems. Though mainly affecting the intestine, other different organs may also be involved like lung, urinary tract or pancreas (1). Majority of NET are seen within the GIT(55%) and respiratory system(30%). Most common site are mainly 45% of small intestine,followed by 20% of rectum, 17% of appendix, 11% of colon and then 7% of stomach(2). High rate of metastases are mainly seen in the aggressive form of Extrapulmonary small cell NE carcinomas(2). According to WHO NE tumor types having following types like atypical carcinoid, typical carcinoid, , small cell lung carcinoma (SCLC), and large cell NE carcinoma (LCNC)(3). Based on their primarily features which are mainly the presence of necrosis and on mitotic counts per 2 sq.mm , WHO criteria for typical carcinoid vs atypical carcinoid vs NEC (3). IHC markers for NET are synaptophysin, chromogranin A,CD56 ,CDX2 and PAX8 are widely accepted and used in pathology lab and practice basis (3) . As dictated by pathology, this type of tumor are differently treated based on their behaviour and aggressiveness (7). The multidirectional differentiation of this concept in which tumors exhibit an combination of neuroendocrine and epithelial cells which begin from a single totipotential cell lead to following classifications distended on the basis of morphologic spectrum of "endocrine tumors" having atypical carcinoid ,carcinoid and small cell carcinoma(8). Here we report an interesting small-cell neuroendocrine carcinoma case of peritoneal mass found incidentally during emergency LSCS.

CASE REPORT

A 27 yr old woman came for emergency LSCS in our hospital. Based on general examination and systemic examination of the central nervous systems respiratory, cardiovascular and abdominal system was normal.Patient was oriented to time, place and person.Peritoneal mass was found during cesarean section and resected.

GROSS

Single, globular , gray-brown to gray- white in appearance, soft tissue piece measuring 6x5x4cm.Outer surface was smooth. Cut surface showed grayish-white solid areas along with few hemorrhagic areas.

Terminology	Differentiation	Grade	Mitotic rates[a] (mitoses/2 mm2)	Ki-67 index[a]
NET, G1	Well differentiated	Low	< 2	< 3%
NET, G2		Intermediate	2-20	3-20%
NET, G3		High	> 20	> 20%
NEC, small cell type (SCNEC)	Poorly differentiated	High	> 20	> 20%
NEC, large cell type (LCNEC)			> 20	> 20%
MINEN	Well or poorly differentiated[c]	Variable[c]	Variable[c]	Variable[c]

Table 1: Classification and Grading Criteria for Neuroendocrine Neoplasms (Nens) of the GI Tract and Hepatopancreatobiliary Organs

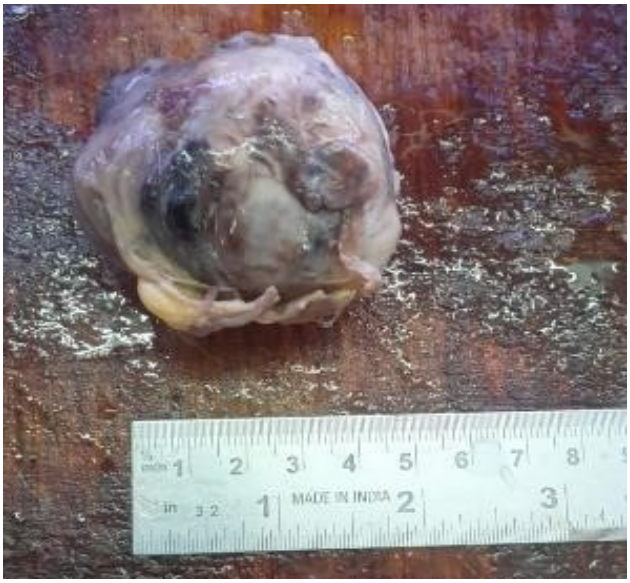

Fig1: Single, Globular, Gray- White to Gray-brown Soft Tissue Piece

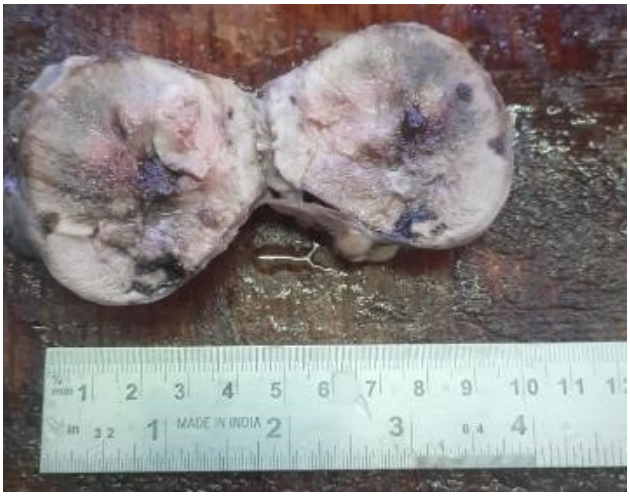

Fig2: Cut Surface Showed Grayish-White Solid areas with few Hemorrhagic areas

MICROSCOPY

Section from the tissue showed partially capsulated tumor comprised of small monotonous tumor cells arranged in trabecular, organoid and acinar pattern. These tumor cells have oval to round nuclei with stippled chromatin -salt and pepper, also surrounded by scant amount of cytoplasm, and Low mitotic counts. Section also showed focal areas of rosette formation along with areas of necrosis and haemorrhage.

DIFFERENTIAL DIAGNOSIS

BASALOID SQUAMOUS CELL CARCINOMA

- Arranged in solid, cribriform, strands and trabeculae of tumor cells.
- Strands of tumor cells often connected to overlying squamous epithelium.
- High mitotic figures.

LYMPHOMA

- large centrocytes,centroblasts,immunoblasts and anaplastic large B lymphoid cells are seen

METASTATIC LUNG CARCINOMA

- Round or oval glands forming acinar gland invading the stroma.
- **In papillary:**

  malignant columnar cells or cuboidal which replace alveolar lining contain fibrovascular cores.

IHC

Synaptophysin and CD56 were positive.

IMPRESSION

Small Cell Neuroendocrine Tumor? Carcinoid

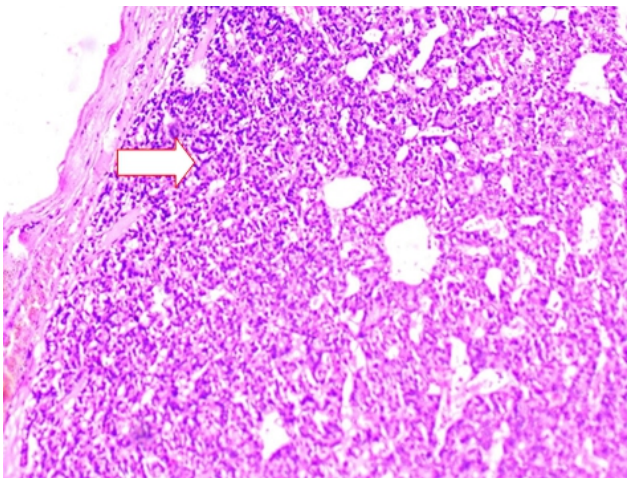

Fig 3: H&E (4X) view of small cell- NET showed small monotonous tumor cells arranged in trabecular, acinar and organoid pattern.

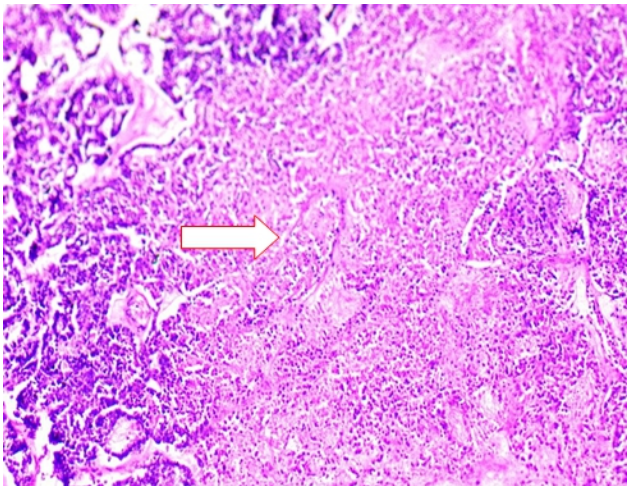

Fig 4: H&E(4X) view of small cell- NET showed focal areas of rosette formation with areas of necrosis and hemorrhage.

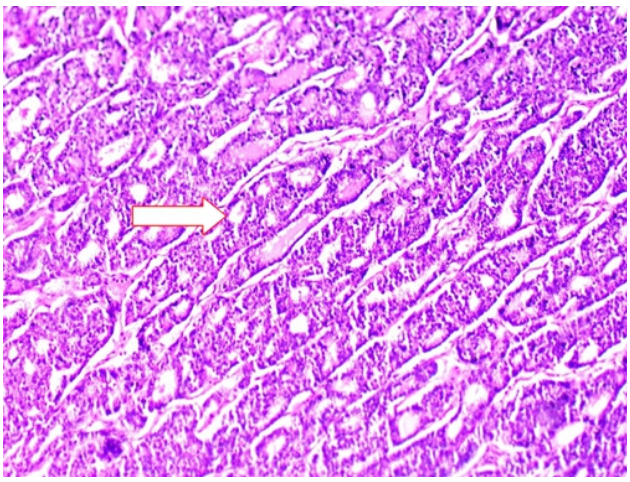

Fig5:H&E (10x)view small cell- NET showed small monotonous tumor cell arranged in trabecular, acinar and organoid pattern.

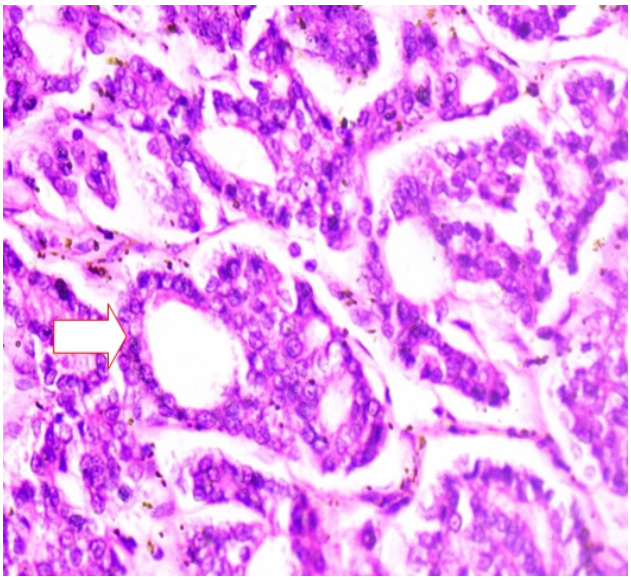

Fig6:H&E(4x)view small cell- NET showed small monotonous tumor cell having oval to round nuclei having salt and pepper chromatin, scant cytoplasm, low mitotic count.

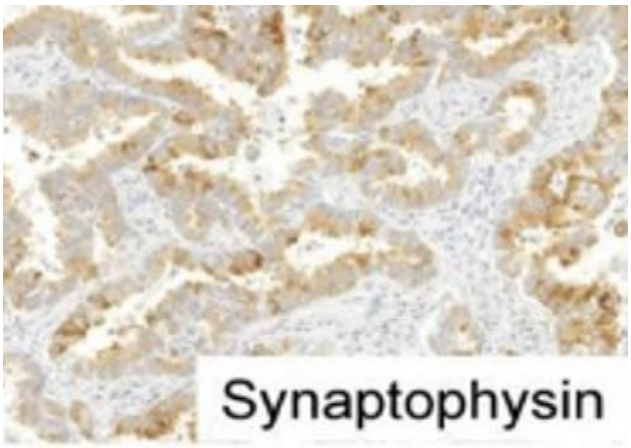

Fig9: A: Synaptophysin showed homogenous moderate to strong positivity.

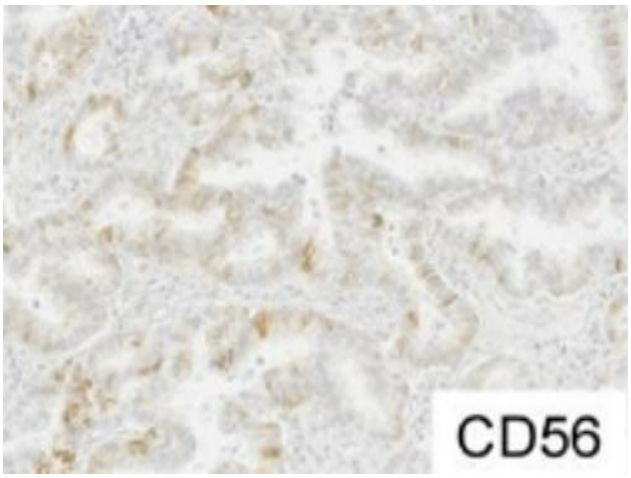

Fig10: B: CD56 Showed focal moderate positivity.

DISCUSSION

According to the WHO , small cell NE carcinoma is histopathologically which are characterized by small size cell , scant amount of cytoplasm, absent or slightly faint nucleoli, granular or stippled chromatin, mitotic rate is high, and also showed frequent areas of necrosis(2).Both pulmonary and extrapulmonary tumors shows identical histologic appearance. Hence, before starting the treatment then it is must be very significant to rule out other causes and sources of many specific primary disease.(2) In our case, due to negative initial type of postoperative imaging for any kind of other faraway lesions, leading to the detection of a primary tumor.(2) Cancer of this type is mainly aggressive, and can occur in younger patients. (2)MRI - the best method to achieve because it gives precise information and clear view images regarding thier location, size, nature and complexity of mass.(2)Neuroendocrine tumour (NET), also known as carcinoid tumour which is rare, slow growing slowly that can occur anywhere throughout the body .(4) Small cell NET is associated with thier early metastases to liver and lymph nodes and associated with poor prognosis(4). Treatment for localized small cell NE carcinoma are mainly done by postoperative chemotherapy or radiation therapy with surgical excision.(6) Radical surgery and adjuvant chemotherapy is the gold standard treatment in cases of metastasis. Immunotherapy has also been proposed and there is continued research to determine the feasibility of this treatment approach(6). On the basis of thier origin, site , mitotic rate count, histopathology proliferation index, and marked by presence of necrosis,it has been classified(7). These type of tumors are mainly identified or specify by their natural history, aggressive behaviour, and widespread metastatic disease on presentation. (7)

CONCLUSION

Neuroendocrine tumours which are very less common which generally originate or arise from the cells of neuro-endocrine system, which may potentially and eventually involve nearly all human tissues. These small neuroendocrine tumor can rarely present as a silent peritoneal mass as seen in the present case. They are highly aggressive tumor having metastatic potential and poor prognosis. Further follow up of the patient is mandatory to rule out metastasis.

REFERENCES

1. Olivieri V, Fortunati V, Bellei Luca, ET AL. Primary small-cell neuroendocrine carcinoma of the bladder: Case report and literature review. Archivio Italiano di Urologia e Andrologia, 92(3)
2. Sisler, K., Knutson, A., McLennan, M. (2019). Primary Small Cell Neuroendocrine Tumor Within a Urethral Diverticulum. Obstetrics & Gynecology, 133(2), 308–311.
3. Natasha Rekhtman.(2022). Lung neuroendocrine neoplasms: recent progress and persistent challenges. 35:36-50.
4. David Ling;Magdalene Chirchir;Nayef Alzahrani;David L. Morris; (2021). Small cell neuroendocrine carcinoma arising in cystic duplication of colon . ANZ Journal of Surgery, (), –. doi:10.1111/ans.17105
5. Al-Ahmadie H, Iyer G. Updates on the genetics and molecular subtypes of urothelial carcinoma and select variants. Surg Pathol Clin. 2018; 11:713-723.
6. Lee DH, Kim JH, Yoon TM, et al. Metastatic small cell neuroendocrine carcinoma of the submandibular gland from the lung: A case report. Medicine . 2020 Jan;99(4):e19018.
7. Niforatos S, Sandhu M, Kallem M, et al. Small Cell Neuroendocrine of the Head and Neck: A Rare Presentation and Review of the Literature. J Investig Med High Impact Case Rep. 2022 Jan-Dec;10
8. Mete O, Wenig BM. Update from the 5th Edition of the World Health Organization Classification of Head and Neck Tumors: Overview of the 2022 WHO Classification of Head and Neck Neuroendocrine Neoplasms. Head Neck Pathol. 2022 Mar;16(1):123-142.

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